CCI 101

2nd Edition

Christopher J. Centeno, M.D.

Dedication: To Ed Jacobsen, Ph.D. for teaching me that the neck can cause more than neck and headache pain.

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Forward and History

Way back when, when I was a young doctor with brown hair (it's grey now) and more enthusiasm than common sense, I had a patient who was different. She had headaches, but also dizziness and brain fog. In looking through the chart, I saw an ENG study by a local audiologist Ed Jacobsen, Ph.D., so I called him and that began an odyssey that ultimately led to a new way to treat CCI.

The audiologist I met that day told me about patients he was seeing for dizziness and imbalance, that his ENG tests told him had neck issues as the cause. I hadn't really heard about anything like that, but after some on-line research (back then you had to have software to search the National Library of medicine), I saw that he was right. This neck and balance connection went way back to the 1920s when two physicians (Barre and Lieou), one French and one Chinese, independently described the syndrome. At the time they thought that this collection of symptoms which included headaches, imbalance, brain fog and other things was caused by damage to the cervical sympathetic chain (posterior cervical sympathetic syndrome). However, what Barre and Lieou syndrome are now credited for is figuring out that headaches and other symptoms can come from the neck.

The next big advance in neck and balance came during NASA research in the 1960s as scientists tried to figure out what damage to the front neck muscles would do to astronauts exposed to levels of acceleration never before experienced by humans. They began to cut the sternocleidomastoid muscles of primates and found that the monkeys lost their balance and would bang into walls.

Then in the 1990s, a new procedure to help neck pain was developed called radiofrequency ablation. The procedure used a radiofrequency probe placed using x-ray guidance to burn the nerves, taking pain from the neck joints and reduce neck pain. However, when the early doctors began to treat the C2-C3 joint in the neck, some patients got permanently dizzy. While later advancements in the technique solved this issue, the upper neck was now firmly implicated in balance.

After confirming what Dr. Jacobsen had told me I soon began treating the upper necks of these patients who had whiplash injuries and also had headaches and dizziness. First with simple muscle trigger point injections and then with upper neck facet injections into the C2-C3 neck joints. Many of them got better. Hence, as more physicians and colleagues learned that this was becoming an area of expertise for our clinic, they referred more of these patients.

However, in this group of patients, we had some people who never got better. When a local chiropractor, Evan Katz came to Boulder and began using a DMX (Digital Motion X-ray), we both began to see that these were those patients who had excessive motion due to damaged upper neck ligaments. Hence, my interest in craniocervical instability (CCI) was born. While more aggressively treating the posterior ligaments helped a bit, most remained largely untreatable.

What was available to these patients? First, many were often bounced from specialist to specialist without answers or any diagnosis. Second, almost all of them would flare up in physical therapy, so many physicians would blame the lack of progress on that patient's lack of effort. Finally, the few that got a diagnosis were just beginning to be offered upper neck fusion, a procedure that in the past would have only been offered to those with neck fractures or severe life-threatening dislocations of the upper neck bones.

Then one day in 2013, I began to play with a model we had of the upper neck bones and ligaments. I literally had it on my desk and would look at it periodically to see if we could access these ligaments. While some of my spinal interventionalist colleagues thought it might be possible to access the ligaments that hold the head on (alar and transverse) from the back, after trying this in a few patients I concluded that it couldn't be done safely as the spinal cord was in the way. It then dawned on me that injections from the front might work as there was a tiny bony tunnel between the C1 and C2 vertebrae.

I then spent months reviewing the anatomy of this area to make sure we wouldn't injure someone and going back and forth with experts on this issue. I tried my first patients in 2015 and by 2016 had enough dialed in that we were starting to see amazing results in these formerly untreatable patients. We added many procedural improvements as the years went on and the rest, as they say, is history.

I've written this book to help CCI patients. My goal is to go over everything they need to know about all of their options. So, I'll cover anatomy, diagnosis, conservative care, surgery, and the procedure we developed. Why take the time to do this as a busy physician? Because patients who know more, in my experience, are the ones that can successfully navigate our medical system to get the best possible results.

However, please realize that in writing this book, I've also tried to "thread the needle" between a patient book and one with enough detail so that patients can give it to their physicians. Why? There is still a serious and devastating lack of knowledge on CCI, and this adversely impacts patients in all sorts of ways. Hence, oftentimes patients need a resource that they can hand to their doctors so that the physician can understand what's wrong with the patient.

Chapter 1-What is CCI?

CCI stands for craniocervical instability. This is the area where the head meets the top of the neck also called the craniocervical junction or CCJ. Instability means that the head or upper neck bones move around too much due to loose ligaments that hold the head in place. Patients often complain of headaches, dizziness, visual problems, and many other problems. We'll get into that all later.

Patients who are ultimately diagnosed with CCI can have a broad swath of disability. I've seen everything from high-functioning patients who can still exercise with symptoms to those who are completely disabled and must lie down most of the day or wear a cervical collar to function.

What Exactly Is Instability?

All of your joints are built to move within certain defined directions. The two things that make that happen are ligaments and muscles. Let's dive in.

Ligaments make sure that your joints can't move too much. When they get damaged, the joint moves in ways in which it wasn't designed. What does this do? It places excessive force on the joint and bones and damages them. In the upper neck, many other structures can get harmed. That includes the facet joints, nerves, cranial nerves, muscles, etc.... As the book progresses, we'll go into all of this anatomy.

CCI Due to Fractures vs. Ligament Laxity

Many surgeons have been taught more about CCI due to upper neck fractures than ligament laxity. These are classical "hangman" type fractures where the upper neck bones are no longer able to protect the spinal cord. This is a surgical emergency as unless something is done to stabilize this area surgically, turning the head could literally cause a spinal cord injury. This type of CCI is VERY RARE.

The type of CCI covered in this book is caused by damaged ligaments and intact bones. This is much more common, but much more mysterious to classically trained spine surgeons. Hence, specialists in this more common type of CCI are few and far between.

The Causes of CCI

Here we can break CCI into two camps: traumatic and congenital. Traumatic is caused by some injury mechanism like a car crash, a manipulation of the head and neck gone bad, or a blow to the head. Congenital means that the patient has a condition that would predispose them to CCI. The most common is EDS, or Ehlers Danlos Syndrome where the patient is born with ligaments that are too stretchy. In that type of CCI, while there can also be an inciting episode, the symptoms can just begin without an event. Traumatic CCI

I've heard many different causes of traumatic CCI that my patients through the decades have relayed:

- Car crash where the head was turned, hit something (like the back of a pick-up truck window), or without head trauma
- Forceful manipulation of the head during a therapeutic manipulation
- Something striking the head
- Falling or inadvertently diving on the head
- A history of old head and neck trauma when younger

There are many more, but these are the most common ones I hear from patients.

One issue to bring up is that some patients report having CCI after being involved in what are classified by insurance companies as low damage crashes. How is this possible?

Cars are designed to deform to absorb force and keep it away from the occupants. However, there are three phases to absorbing forces. The first is, as you see above, when the car is elastic (not much damage), since the car is not absorbing force, it gets transferred to the occupant. This is where many patients get injured. Next, in crashes where the car starts to deform, the force on the occupant goes down because now the car is now absorbing the brunt of the force. Finally, in crashes where big damage happens, you get immense forces on the occupant. My friend and college Michael Freeman, a forensic epidemiologist recently published an excellent paper that quantified why the insurance company position that patients can't be injured in a low damage crash is not supported by crash test data (26).

Congenital CCI

When a patient is born with ligaments that are too stretchy, this condition is known as Ehlers Danlos Syndrome or EDS. In this condition, the body makes collagen with the wrong mix of components. This is a spectrum disorder from milder ligament laxity to more severe. As patients with more severe ligament laxity get older, all of the extra wear and tear and accumulated injuries over time begin to cause pain and disability. One such area where some patients' manifest problems is the cranio-cervical junction.

Because of the overly stretchy ligaments that hold the head on, EDS patients have a lower threshold for injury here. So, while all of the above listed traumas can cause CCI, lesser things like banging the head on a car while getting out can cause it as well. Other times there is no specific trauma that these patients can point to, the CCI symptoms just begin to manifest.

Chapter 2-Getting to a Diagnosis

The diagnostic journey for CCI is often long and arduous. Few patients get to the right specialists quickly and get a concrete diagnosis within months of the onset of symptoms. In fact, many end up going down unfruitful and costly rabbit holes as the medical care system focuses on only one symptom and works that up. So, for example, here are common unfruitful journeys for CCI patients based on their symptoms:

- Headaches that are caused by the CCI end up getting worked up and treated by a neurologist as migraines which are a different animal involving blood vessels in the brain.
- Dizziness/Imbalance/Lightheadedness caused by the bad position sense information coming in from the damaged upper neck is often worked up as a vestibular problem by an ENT doctor with tests like a VNG.
- Rapid heart rate caused by Vagus nerve irritation is often worked up by a cardiologist with a Holter or other wearable monitor to rule out supraventricular tachycardia.
- Brain fog or problems concentrating caused by the weird inputs coming in from the damaged neck is often worked up by a neurologist as a brain injury, stroke, or a seizure disorder.

We use many different things to get to a CCI diagnosis. I'll cover these here:

- History of Onset-see above in causes of CCI section
- Symptoms
- Response to treatment
- Imaging

The Symptoms of CCI

There has never been a formal published survey of CCI symptoms. Hence, I compiled the symptoms of 80 patients who we confirmed as having CCI and who were about to undergo a PICL procedure. This was the word cloud of what they reported:

The bigger the word, the more often it was reported. Based on this data and decades of experience treating these patients, this is how symptoms break down.

These are classical symptoms that almost all CCI patients would have like:

- Headaches
- Upper neck pain
- Dizziness/Imbalance/lightheadedness
- Brain fog
- Clicking/popping in the neck (crepitus)

Then there is a collection of symptoms shared by most CCI but not all patients:

- Ringing in ears (tinnitus)
- Shoulder pain
- Rapid heart rate (tachycardia)-POTS and/or increased anxiety
- TMJ
- Memory Loss
- Wandering pain or numbness/tingling in various areas of the body
- Gastric disturbances
- Clicking or popping in the upper neck with movement
- Fatigue
- Nausea

Then there is a collection of symptoms shared by some CCI patients:

- Facial pain
- Tongue symptoms
- Loss of consciousness episodes
- Sweating

Hence, diagnosing CCI by symptoms alone can be very difficult.

A Word of Advice on Symptoms

As you can see, CCI patients can have a large number of symptoms. During this book I'll go through various rabbit holes of misdiagnosis to avoid. One tip here is that patients these days often find like patients on social media and begin self-diagnosing. When they see a doctor, they begin using more diagnostic terms like "atypical tachycardia" rather than just saying that when my neck is bad my heart races. This can often confuse the diagnosis for the doctor. Another thing they do is to begin listing every possible symptom they believe is related to their CCI rather than just the main symptoms they know are for sure related. This also can make the doctor confused. So, when you talk to a doctor about your problem, keep it focused on what you experience and keep it streamlined and simple.

Response to Treatment

CCI patients have a few classical responses to treatment. One is that active physical therapy focused on strengthening tends to make them worse. Why? The upper neck ligaments are just too loose. Hence, when they try to strengthen the neck muscles, the upper neck moves around too much and irritates joints, nerves, tendons, etc....

On the other hand, many patients find good temporary relief from upper cervical chiropractic adjustments (NUCCA or AO chiropractic). They have improved symptoms that can last for hours to weeks. This can also be highly dependent on finding the right chiropractic expert. Meaning not all upper cervical chiropractors have success in all CCI patients and some patients find only a handful of expert chiros who can handle their case.

Some quick definitions here:

- NUCCA (National Upper Cervical Chiropractic Association) Chiropractic-focused on gentle manual adjustments of the upper neck bones that are guided through precise x-ray measurements.
- AO (Atlas Orthogonal) Chiropractic-Very similar to NUCCA, but the x-rays are used to program a vibration device which performs the adjustment.
- Blair Chiropractic-Also focused on gentle manual adjustments of the upper neck as well as diagnostic imaging where angles are calculated to drive what needs to be adjusted. There is also a leg length test that's commonly used to see how the upper neck is impacting the rest of the body, all the way down where the feet meet the ground.

Other types of treatments where we look for a response are injections. For example, upper neck instability can cause the upper neck facet joints to get beat up and hurt. Hence, in many CCI patients, injecting the C0-C3 facet joints will help their pain. Or for example, occipital nerve blocks which can help headache pain. The focus here is to determine if there are structures in the upper neck causing pain as that helps the doctor pinpoint the area of the problem.

Imaging

There are many ways to try to image the problem of CCI. However, before we get into that in chapter 3, we need to learn a little anatomy.

Examples of How These Pieces Come Together

You now have the diagnostic categories, which to review are:

- History of Onset-see above in causes of CCI section
- Symptoms
- Response to treatment
- Imaging

So now let's review some examples. While we still have some things to learn as we move forward, this is just to show you how a CCI diagnosis would be made.

Example 1-A patient was in a car crash where they were rear ended while their head was turned. Now let's plus some things in:

- History of Onset-The problem began at the car crash, which was sufficient to cause this kind of injury.
- Symptoms-Headache, dizziness, brain fog, and upper neck pain. Hence the symptoms match CCI.

- Response to treatment-Physical therapy makes the symptoms worse. An upper cervical facet injection decreased headache pain for 3 weeks. Both of these things fit the profile of how a CCI patient would respond to treatment.
- Imaging-The patient's DMX study shows 6 mm of overhang which fits with instability.

All of this shows that we're 4/4 in determining if this patient has CCI.

Example 2-A patient who has EDS bangs their head on the roof of a cab while getting out. Here's the analysis:

- History of Onset-The patient has EDS, which means that they have a major risk factor for CCI. The new problems began getting out of the car, which could injure the upper neck ligaments in an EDS patient who already has stretchy ligaments.
- Symptoms-Headache, neck pain, rapid heart rate, GI problems, and brain fog. Hence, the symptoms match.
- Response to treatment-Upper cervical chiropractic (NUCCA) helps the symptoms for days at a time, but then they return. Home strengthening makes the patient worse. Both of these responses to treatment match what a typical CCI patient would report.
- Imaging-The patient's flexion-extension MRI shows that their Grabb-Oakes measurement increase from 9 mm to 12 mm with flexion. This matches CCI.

Again, another 4/4 match.

Example 3-Now we have a patient who has no history of trauma whose symptoms just began without any warning:

- History of Onset-No trauma, no risk factors for CCI.
- Symptoms-Brain fog, rapid heart rate, GI problems, and jaw pain. This may or may not be a match.
- Response to treatment-Upper cervical chiropractic (NUCCA) doesn't help the symptoms. Strengthening is not a problem; it also doesn't help. This doesn't match CCI.
- Imaging-There are no concrete measurements that match CCI other than one measurement which is borderline.

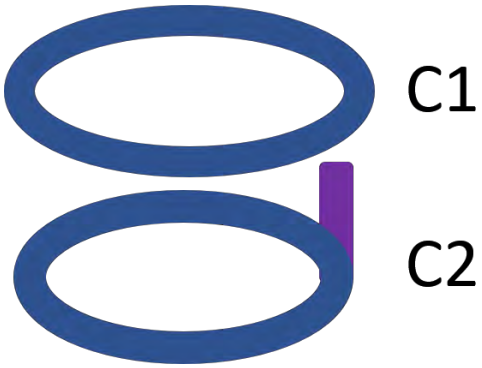
So, we have a 1.5/4 here, so this is less likely to be CCI.

Chapter 3-Upper Cervical Anatomy and Basic CCI Concepts

While this next chapter may seem tedious, it's essential for anyone with CCI to understand how the basic anatomy works. I've purposefully made this chapter a bit more complex that the average patient may want to know, to again, thread that needle between a book that can help patients versus one that's detailed enough to also help their doctors understand CCI. If it gets too intense for you as a patient, then read as much as you want to know and skip forward to the next chapter.

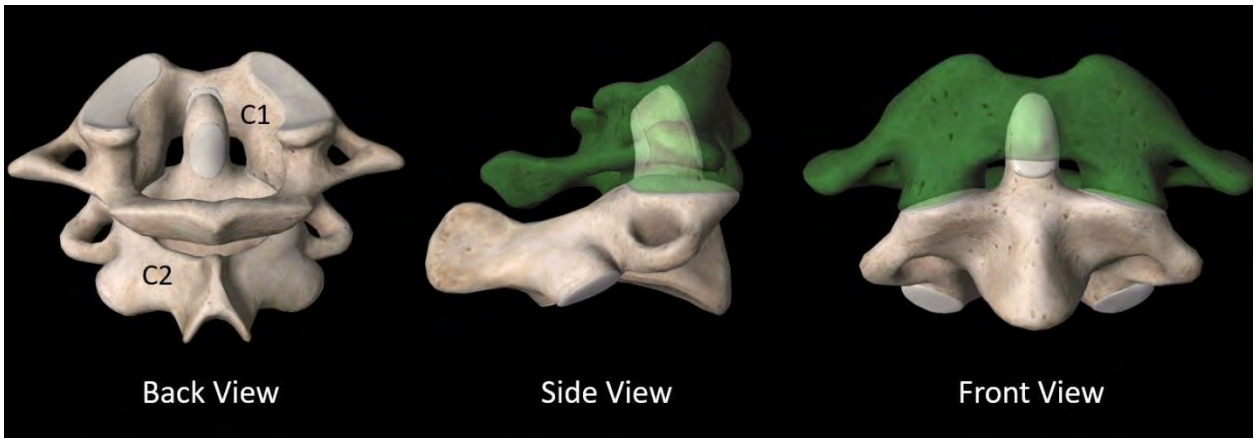
Bones

The head on the neck is like a bowling ball on a stick, meaning it's inherently unstable. Hence, we have two things that evolved to hold it on. Tough ligaments and muscles. In addition, the upper neck bones are specially adapted to help this effort as well. Let us begin there.

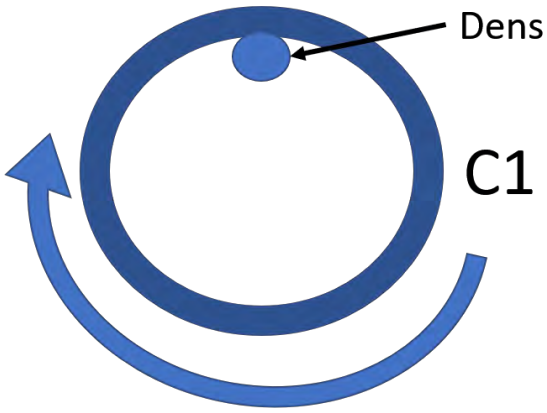


The neck bones are numbered. C0 is the skull and C1 is called the atlas while C2 is called the axis. Both are ring shaped bones that I have simplified above. The peg of C2 fits into C1 and the skull rests upon C1. There are also joints connecting the skull to C1 and C1 and C2 (not shown here). These are called facet joints.

Here's what the bones really look like:

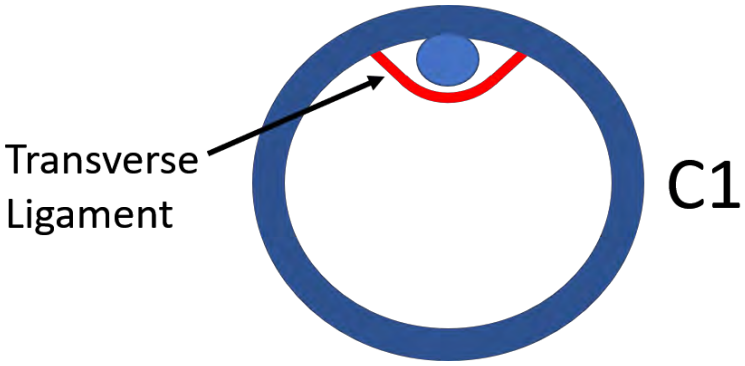


Now back to our simplified drawings. The peg in C2 (Dens) is there so the head and the atlas can rotate around it:



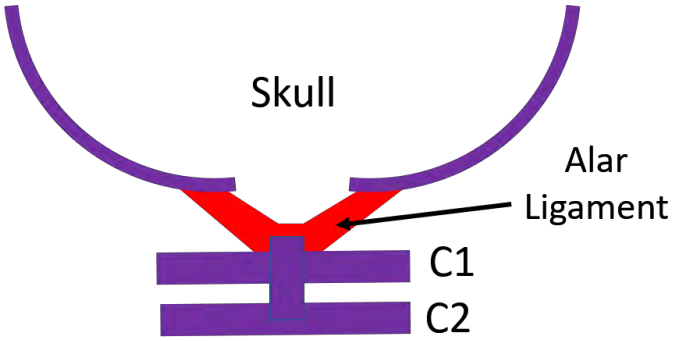
Ligaments

How would we keep these two bones together? That's where ligaments start factoring in. The big one here is the transverse ligament (aka the transverse band of the cruciform ligament):

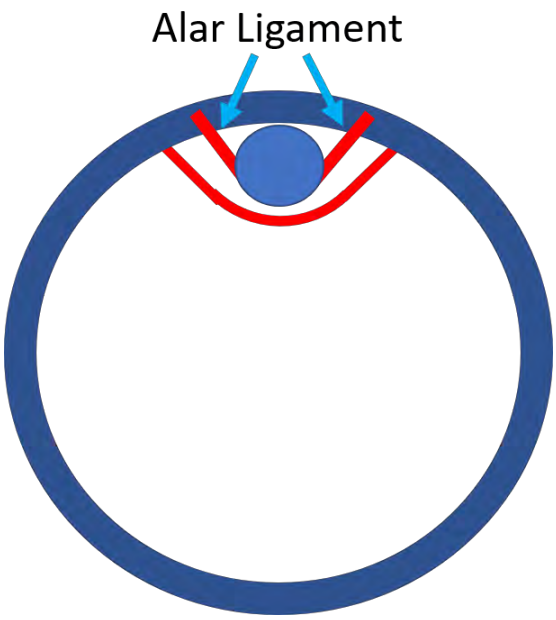


If this transverse ligament is loose, it allows too much movement of C1 on C2 when the head is flexed forward.

However, how do we keep these two bones firmly attached to the skull? The alar ligament does that job:

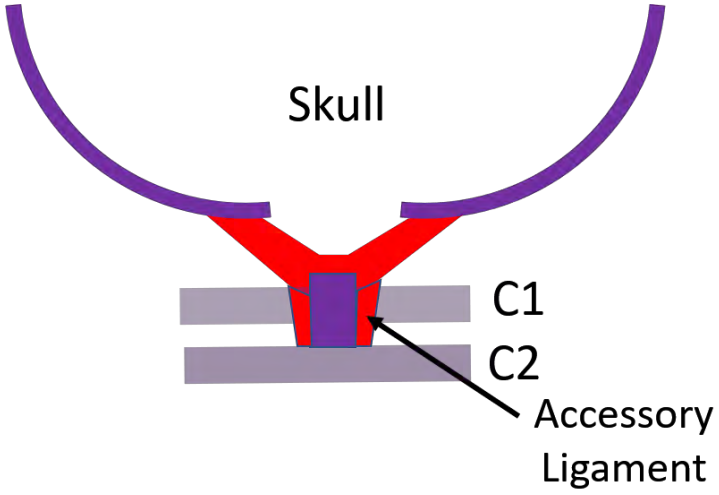


This ligament goes from the dens to the skull base, kind of like arms reaching up from the dens to the skull. Hence, it binds C2 to the skull and indirectly holds C1 in place as well by sandwiching it between C2 and the skull. When this ligament is loose or damaged, it can allow C1 to rotate on C2 because it no longer applies that sandwich force to help keep C1 in place. In addition, when damaged, it can also allow the skull and C1 to slide sideways when the head is side bent. We measure this as C1-C2 "over-hang" on a DMX study (more on that later).

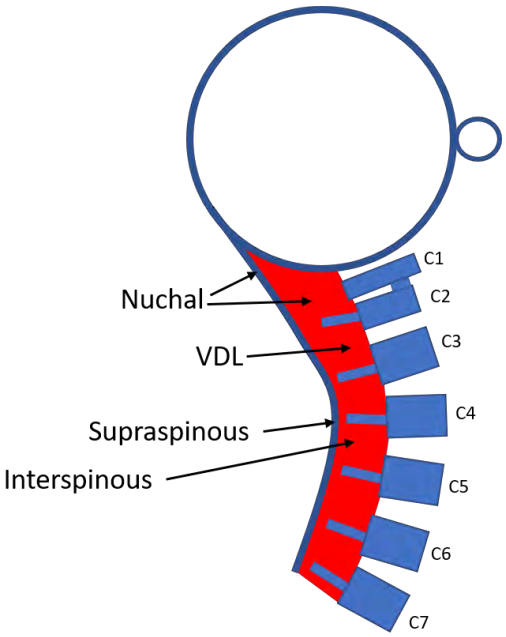


It should also be noted that when viewed above, one band of the alar ligament travels toward the front as shown here. Remember that this ligament goes to the base of the skull. Hence, then the head and neck bend forward, the weight of the head is partially supported by this ligament.

There's another part of the alar ligament which extends downward between the alar ligament and C2 which is called the accessory ligament as shown below (in this picture I made the C1 and C2 bones transparent (except for the dens)):

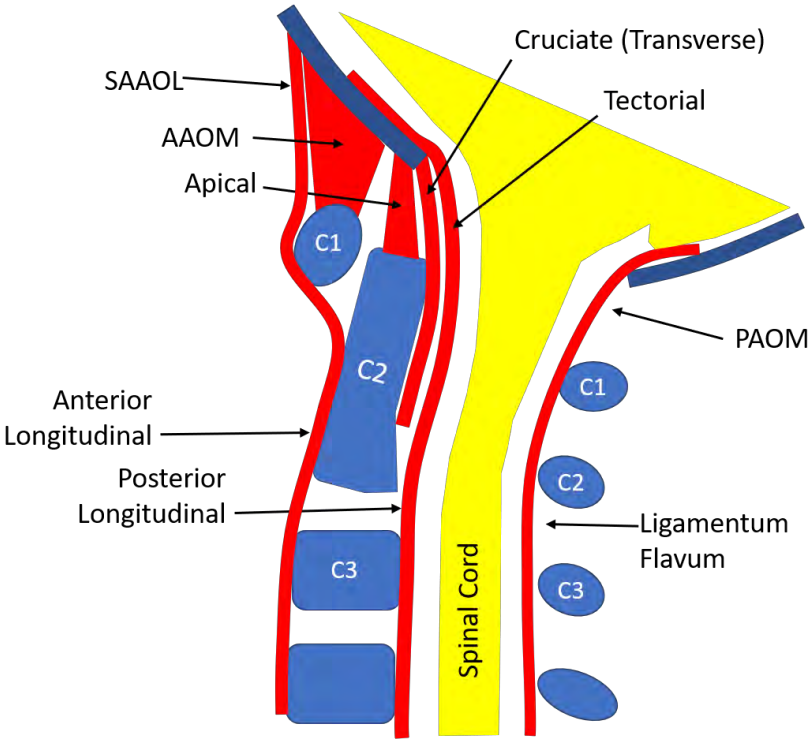


If we look from the side at the whole neck, we see that there are lots of ligaments in the back or posterior part:



These are the nuchal, VDL (Vertebro-dural Ligament), supraspinous, and interspinous. They all help prevent the vertebrae from moving too far forward and are very active when you look down. The VDL is special because it connects directly to the dura, which is the covering of the brain and spinal cord. Hence, it is believed that damage to this ligament may cause headaches.

Finally, there are deeper cervical ligaments to consider, which are shown below:



These include from front to back:

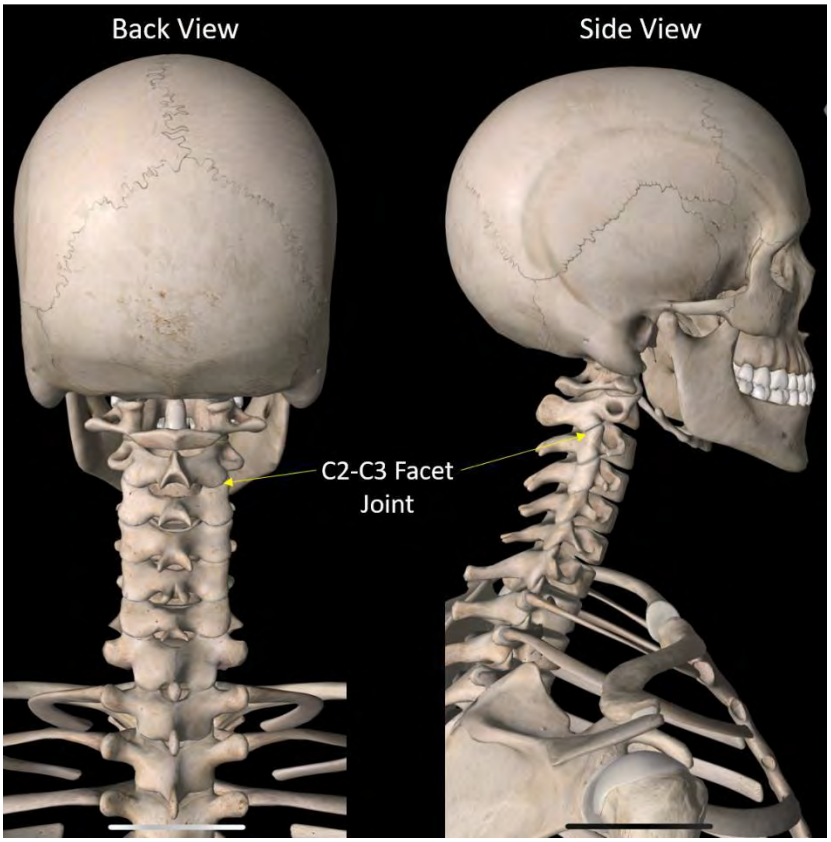
- SAAOL-Superficial Anterior Atlantooccipital Ligament (this is the extension of the anterior longitudinal ligament upwards. This connects the front of the cervical vertebral column with the skull.
- AAOM-Anterior Atlantooccipital Membrane-This connects C1 to the skull.
- Apical-This extends from the dens of C2 to the skull.
- Cruciate-This is a cross shaped ligament that has as its horizontal part the transverse ligament discussed above but has parts that go up and down as well.
- Tectorial-This is the upward extension of the posterior longitudinal ligament that connects the back of the vertebral column to the skull.
- PAOM-This is the upward extension of the ligamentum flavum that connects the back part of the spinal canal to the skull.

The front ligaments help to stabilize your head on your neck when you look up, the middle ones help to hold your head on, and the back ligaments keep everything stable when you look down.

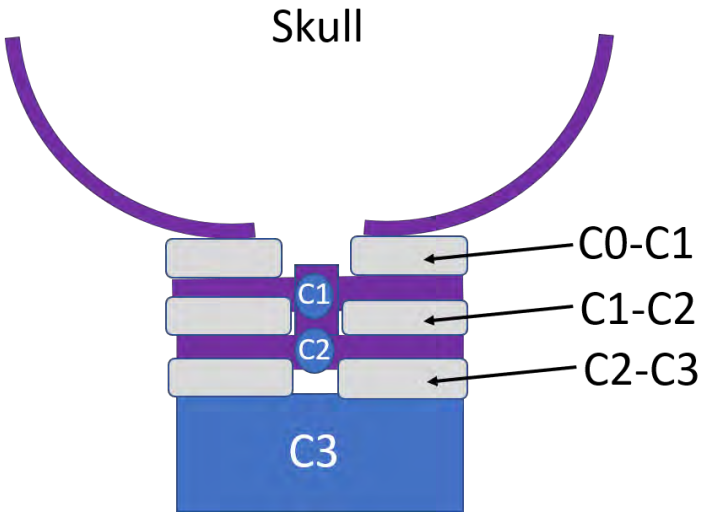
Facet Joints

Your upper neck (and the rest of it as well), has finger size joints called facets. Their purpose is to help your neck move normally. When the ligaments are damaged, the joints can sustain too much wear and tear and begin to get injured.

This is how those joints really look:



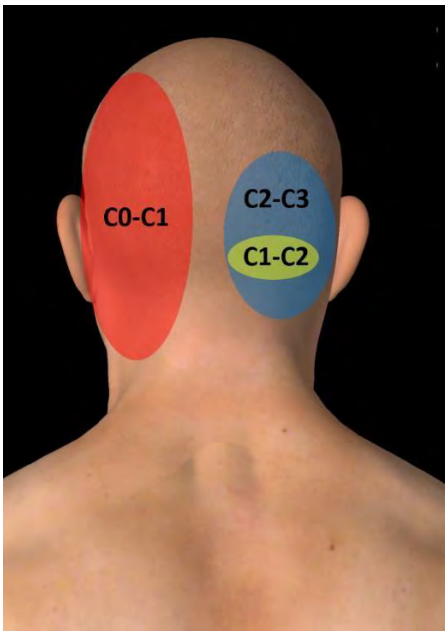
Here's a simplified diagram:



These upper neck joints are

- C0-C1-This joint allows the nodding movement of the head. If injured or painful, this joint can cause pain in the back of the head.
- C1-C2-This joint allows for about half of the rotation of your head. This joint can cause pain to the back of your upper neck where your head and neck meet.
- C2-C3-This joint allows for a little rotation of the head and a little forward bending of the neck. It causes pain in the upper neck and the back of the head.

These are the referred pain patterns for these joints (24):

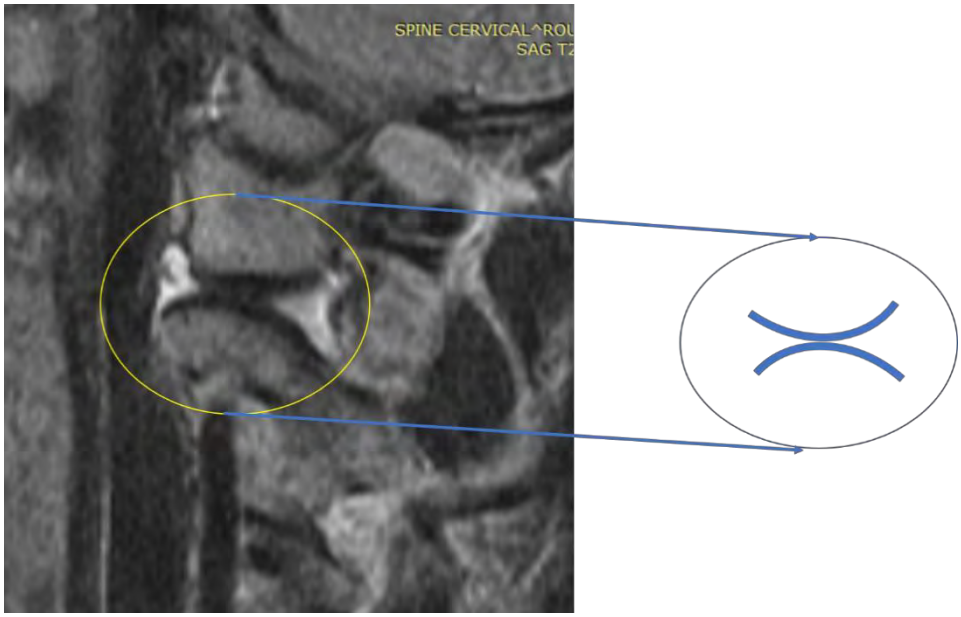


The red area is where pain from the C0-C1 joints is felt. The yellow/green is where pain from the C1-C2 joint localizes. Finally, the blue area is where pain coming from the C2-C3 joint travels.

Hence, these upper neck joints can cause headaches. In addition, all of them also provide information to the brain about head, neck, and eye position. This happens through tiny sensors present in the joint capsules called proprioceptors. Hence, if these joints are injured, they can cause dizziness, imbalance, or visual problems.

C1-C2 and Inherent Instability

The C1-C2 facet joint is responsible for 50% of the total neck rotation. Like all facet joints, it has cartilage and a joint capsule. What makes this joint very unique is the shape of its internal joint surfaces:



Above is an MRI side view of the C1-C2 facet joint. Its surfaces are shown to the right. Note that they are both convex, which is unusual in the body. Why? Usually, the joint surfaces fit together like a puzzle piece with one surface being convex and the other being concave. That makes them inherently stable. However, the C1-C2 joint is inherently unstable.

What does that mean? If we have a convex/concave joint like the one below and there are no muscles or ligaments acting on it, it's not going anywhere. Meaning the joint surfaces stay together. The opposite is true if the joint surfaces are convex sitting on top of convex:

Convex/Concave Joint Surfaces-Stable

Convex/Convex Joint Surfaces-Unstable

So, the C1-C2 joint is inherently unstable. This explains why we have an entire field of chiropractic care devoted to this part of the neck! However, the C0-C1 joint above it is that convex/concave type, so it is inherently more stable.

In the section above, we discussed how the atlas (C1) was sandwiched between the skull and C2 and held in just the right amount of tension by the alar ligaments. These ligaments help to maintain the concave joint surfaces in approximation as you move your neck. However, if this ligament is damaged, these joint surfaces cause the bones to rotate (25).

Why Inherent C1-C2 Instability Can Wreak Havoc with Patients

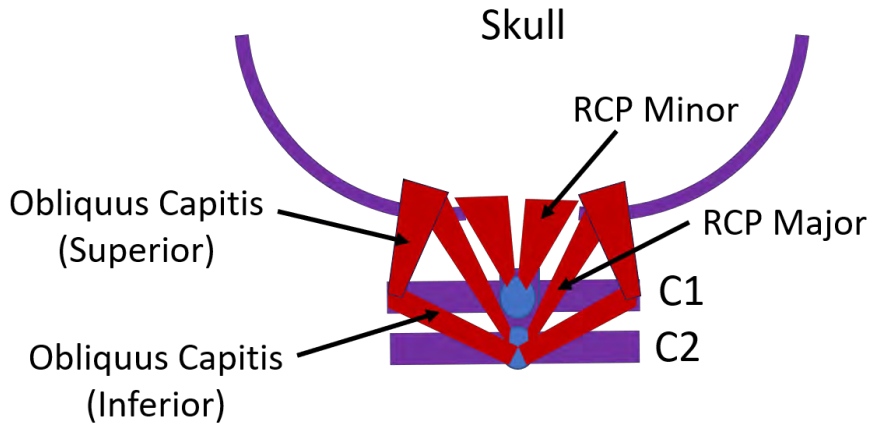
Given that the C1-C2 joint is inherently unstable, if it doesn't have ligaments and muscles to hold it in place, it tends to move out of place. This results in the C1 bone rotating on the C2. In fact, this is why an entire type of chiropractic (NUCCA) is devoted to trying to get C1-C2 back in place. So, if you have C1-C2 instability due to damaged ligaments (Craniocervical Instability or CCI) it's easy to see why this joint would not be "in place".

What happens when this joint gets out of whack? The capsule is rich in position sensors that help maintain the position of the body relative to the neck, so your body will feel out of whack as well. In addition, the joint, when painful, refers its pain to the head. So, you can also have headaches and other symptoms (see below).

In addition, it's possible for this joint to be out of whack without having damaged ligaments. This is where a NUCCA chiropractor or an experienced manual physical therapist can make a big difference. Symptoms here include headaches, dizziness/imbalance, visual disturbances, brain fog, or spasm, or pain at the back of the head.

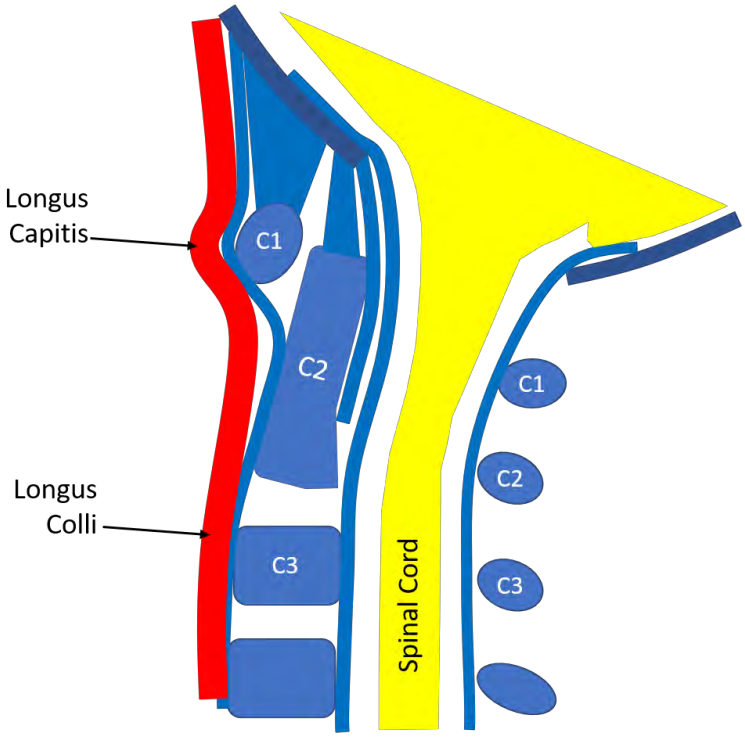
Muscles

The muscles in this area all control the fine movements of bowling ball in the stick:



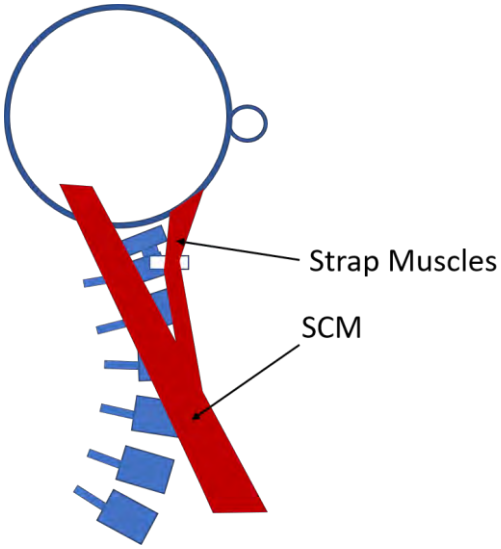
The RCP (Rectus Capitis Posterior) major and minor connect the C2 and C1 muscles to the skull and can help you look up. The RCP minor is unique in that it connects directly to the covering of the brain and spinal cord (dura), so it's implicated in headaches. The obliquus capitis muscles connect C1 to C1 and the skull and help with bending your head sideways on your neck.

The deep muscles in the front of the neck look like this:



The longus capitis and colli live just in front of the spinal column and help to flex the neck and maintain the curve.

As we move outward, we find the strap muscles in front including the SCM:

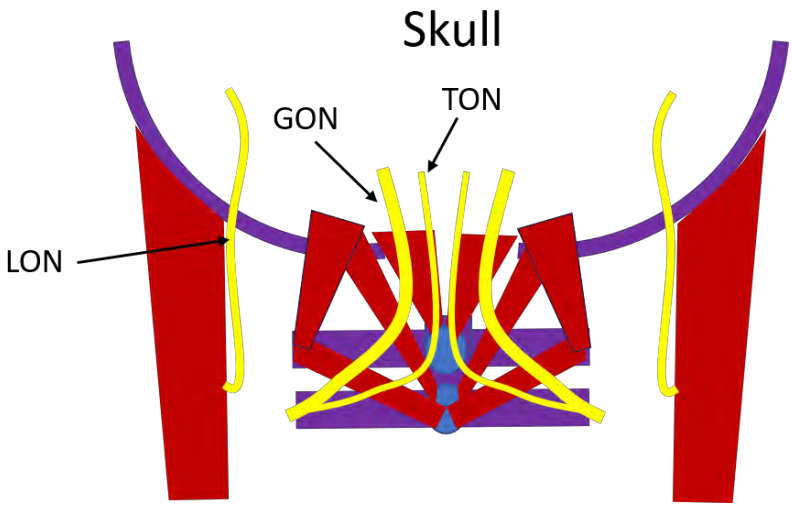


The SCM is a critical muscle up front as it is the prime mover for rotating the head. This is important for CCI as C1-C2 is a critical joint that's responsible for 50% of your ability to rotate.

There is a slew of other muscles like the upper trapezius, levator scapula, and paraspinals muscles that usually get in on the act in CCI, but we'll discuss those later.

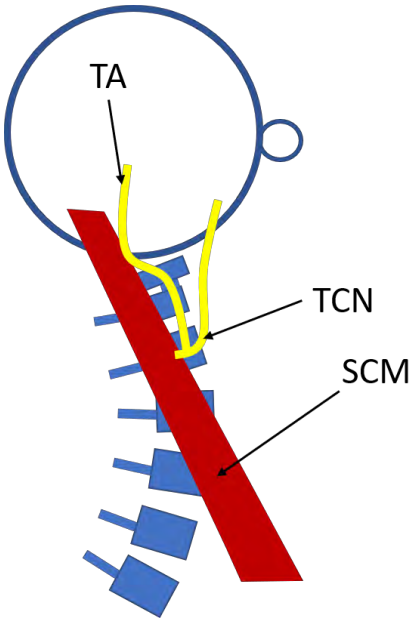
Nerves

Pissed off upper neck nerves tend to cause headaches. The nerves that are critical for CCI live in both the back and front of the neck. Here are the ones in the back of the neck:



Realize that since these upper neck muscles help to hold and stabilize the head, when they spasm and go into overdrive to help stabile the head on the neck, they can irritate these nerves and can cause headaches. There are also nerves that live and exit in front of and behind the SCM:

- TON-Third Occipital Nerve
- GON-Greater Occipital Nerve
- LON-Lesser Occipital Nerve

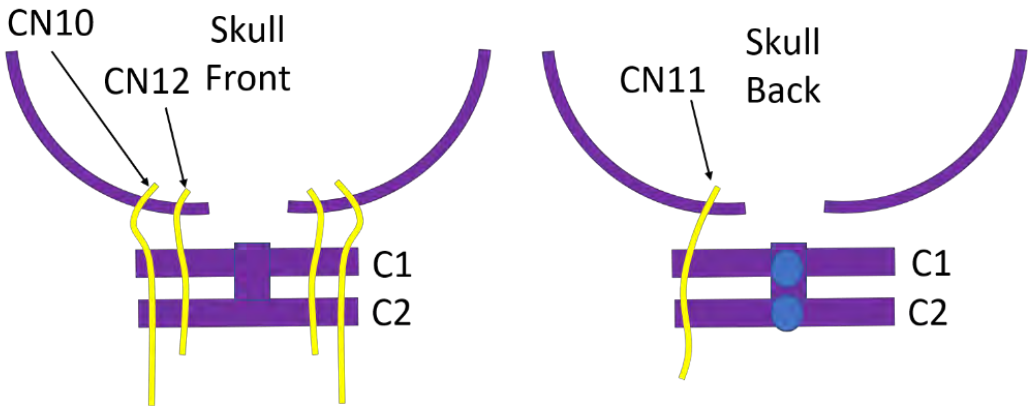


The TCN is the transverse cervical nerve (also called the superficial cervical plexus) and the TA is the temporal-auricular nerve (also called auriculo-temporal nerve). The TCN nerves live behind the SCM

muscles, so they can become irritated as those muscles get tight in C1-C2 instability. They can cause headaches, as well as jaw, side of head and face pain. Note that the TA nerve also travels right behind the jaw, so any extra motion or overload of the jaw can cause it to be irritated and this can lead to headaches at the side of the head. you

Cranial Nerves

The cranial nerves come out the skull and head south, as shown below:

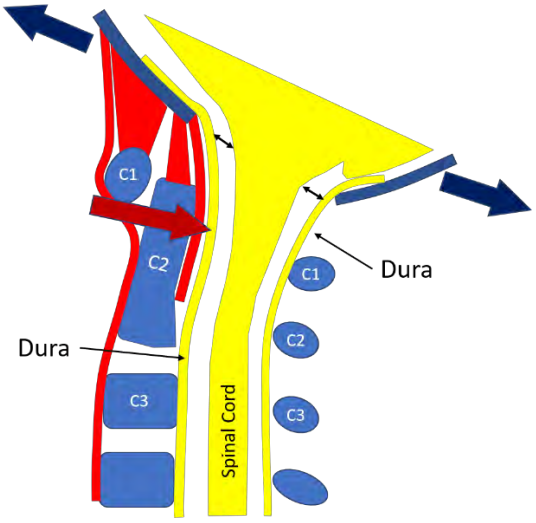


Given that the skull moves too much on the upper neck and these nerves also are close to the atlas bone which can move around too much as well, these nerves can get irritated. The more common cranial nerves are:

- CN10-Vagus Nerve-This is literally the brakes on the heart, lungs, and digestive tract. If the nerve gets irritated the patient can get rapid heart rate, anxiety, and digestive problems.
- CN11-Spinal Accessory Nerve-Controls the upper trapezius and sternocleidomastoid (SCM) muscles. When it gets irritated these muscles can go into spasm.
- CN12-Cranial Nerve 12-Hypoglossal-Controls the tongue muscles. This can cause spasms in the tongue area.
- CN V and VII-These both go to the face.

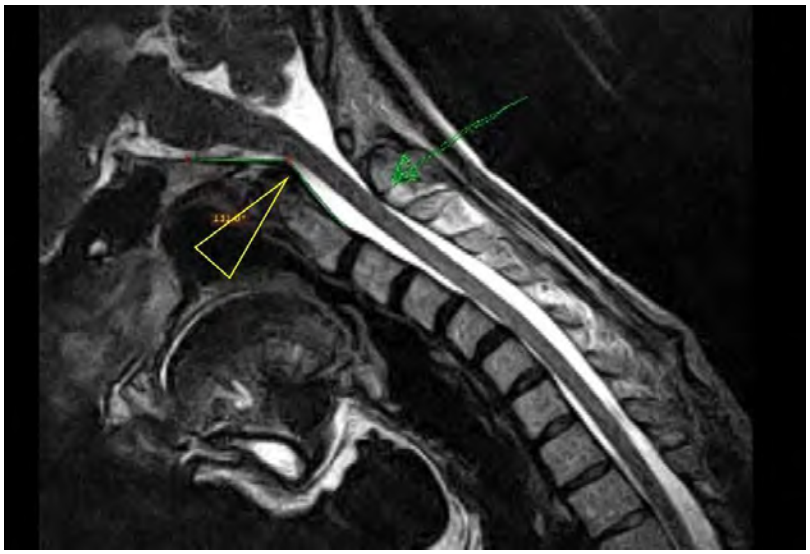
Foramen Magnum

The base of the skull has a big hole in it and the name for that in Latin is “foramen magnum”, which is what we doctors call it. Realize that this area has quite a bit of room for the spinal cord, which is great as that means that quite a bit of motion can happen here before there is a spinal cord injury:



The black arrows on either side of the yellow spinal cord show that room, but also note the yellow line which is the covering of the spinal cord and brain (the dura) which is closely adhered to ligaments. So, if that area gets irritated due to CCI (too much skull movement the dark blue arrows), then while there is no spinal cord injury, there can be all sorts of referred pain in all sorts of different places in the body. In addition, in many CCI patients the dens (here marked C2) can move backwards due to a loose transverse ligament, which can cause pressure on the dura in the front and possibly the spinal cord (also called cervical medullary syndrome).

Cervical Medullary Syndrome



As discussed above, when there's instability, the skull moves too much against the spine and as a result, the dens can move too far backward (yellow arrowhead) and irritate the top part of the spinal cord or brainstem (medulla). When that happens, as shown above, that's called cervical medullary syndrome.

Also realize that the white stuff you see above on either side of the spinal cord is cerebrospinal fluid (CSF). This circulates around the brain and acts as the waste removal system for chemicals produced in the brain. This circulates all the way down to the neck, upper back, and lower back. The flow of that fluid

can be obstructed by a cervical medullary syndrome. We'll go into more detail later on how that's measured using specialized MRI.

This is where the concept of a pannus on the back of the dens comes in. A pannus in the context of CCI is swelling and sometimes scar tissue in the dens bursa. This can further close the space between the dens and the upper cervical spinal cord and medulla, leading to more potential compression of these structures. Having said that, it's critical to note that 90% of CCI patients that have a pannus do not need surgery to remove it, they just need to get the CCI treated and stabilized through injections to reduce the swelling leading to the pannus.

Types of CCI

The type of CCI you have dictates the treatment you need. I can't stress this enough. As a result, I have created a CCI sub-typing system that is broken down by upper cervical level:

- Type 1 CCI involves the C0-C1
- Type 2 CCI corresponds to the C1-C2
- Type 3 CCI is focused on the C2-C3

Each of these levels is broken down further based on which ligaments are problematic and which measurements are abnormal.

Type 1 CCI:

This type is broken down into five categories:

- Type 1a-Lax middle and posterior ligaments like Tectorial, PAOM, and nuchal ligaments-Low CXA in flexion
- Type 1b-High CXA in extension-Lax anterior ligaments like AAOM, sAAOL, anterior C0-C1 facet capsules
- Type 1c-Translational Instability at C0-C1-Shallow occipital condyles along with other ligaments being lax like the C0-C1 facet capsules and some of the ligaments involved in Types 1a and 1b.
- Type 1d-Rotational instability at C0-C1-Lax alar, C0-C1 facet capsules, and anterior-posterior atlanto-axial ligaments. Excessive C0-C1 rotation on movement-based CT or MRI.
- Type 1e-Vertical Instability at C0-C1-Lax apical dens, alar, AAOM, and PAOM ligaments with the skull moving too far vertically upward. Basion-Dens index is too high in neutral, flexion, or extension on upright MRI.

Type 2 CCI:

- Type 2a-High ADI in flexion-Transverse ligament laxity. High ADI in flexion.
- Type 2b-Excessive C1-C2 overhang in APOM lateral bending view. Alar ligament laxity with excessive C1-C2 lateral translation with lateral bending open mouth view on DMX.

Type 3 CCI:

- Type 3a-Translation in flexion at C2-C3-Lax supraspinous/interspinous ligaments with excessive translation in flexion.
- Type 3b-Translation in Extension at C2-C3-Lax ALL ligament with excessive translation in extension.

Craniocervical Syndrome vs CCI

Can someone have all of the same symptoms as CCI but not instability? The answer is a resounding yes and we call that craniocervical syndrome or CCS.

Remember that most of the symptoms of CCI are generated from the upper neck structures. For example, dizziness can be caused by abnormal position sense information being generated by the upper neck facet joints. So, if these joints are irritated or injured without the ligaments failing, you get dizziness without the instability or what we call CCS. Headaches can be caused by irritation of these joints or the occipital nerves, so again, you can get headaches without instability.

Treatment for CCS is simply targeting the same structures we would normally treat in CCI patients without needing to worry about targeting the ligaments.

Adjacent Segment Disease and CCI

Another thing to mention is how the lower neck or other areas of the spine can impact the upper neck. Each segment in your spine is meant to move in a balanced left/right and front/back motion. So, what would happen if we stopped a part of your spine from moving or lost that balance? With respect to your CCI, we call that Adjacent Segment Disease or ASD.

ASD means that the levels above or below are getting abnormal forces and as a result, are causing problems. An easy to conceptualize example is if a patient has a fusion. This means that one or more levels are bolted together so they don't move at all. Hence, the levels above and below can develop ASD. Why? The forces that would normally be absorbed by the fused levels can no longer be handled by those levels and therefore get distributed above and below.

ASD can be a problem for CCI patients as many also have problems in their lower spine. For example, patients with large disc bulges at C4-C5, C5-C6, or C6-C7. Many of these patients end up getting fused at these levels because that's the "bright shiny object" on MRI. Regrettably for some, these levels weren't causing their symptoms, but instead those symptoms were being generated by the upper neck and CCI. Hence, when they get these lower neck levels fused, the CCI gets worse because more forces are pushed up to those upper levels by the lower neck fusion. We also see patients who have a legitimate issue in the lower neck causing some of their symptoms like numbness and tingling in one or more hands. These symptoms resolve after lower neck surgical fusion, but then the upper neck symptoms worsen.

ASD can also happen due to scoliosis, and this should be separated out from CCI. How does that work? Remember that scoliosis is a side bending of the spine. Any time the spine side bends, it also rotates. This means that in scoliosis patients, there is a rotational force imparted on the upper neck. Meaning, scoliosis forces C1 to rotate on C2. This can cause CCI like symptoms and over time can even stretch out various ligaments to cause a fixed rotation of those segments.

How can you help ASD patients with CCI? First, hopefully we can intervene before the fusion. To this end, we can use the same technologies we use to help CCI to reduce the symptoms or help the lower neck. If the fusion has to be done, then we may be chasing those extra forces and the patient may get a less than permanent result, but we can still usually help. If the patient has scoliosis, then this can also be treated with regenerative medicine type therapies, but the twisting forces in the upper neck may again need to be chased rather than permanently fixed. In addition, physical therapy types like Scroth which help with left/right balance in the spine may be helpful.

Chapter 4-Imaging

Imaging in medicine means that some technology is used to visualize the deep structures of the body. This is always an interesting topic with CCI patients. What I mean is that this is such a huge topic that it's tough to cover all of it. In the meantime, many CCI patients see surgeons who use some of the metrics I'll cover here to determine if they're surgical candidates, so I want to make sure to cover those measurements. However, to make this part of the blook less cluttered, I'll give brief descriptions of the measurements and then point to a YouTube video or reference that goes into more depth. **Imaging 101**

X-ray Exposure

It's important to understand a bit about common types of imaging discussed here and x-ray exposure. For example, a simple neck x-ray has about the same amount of x-ray exposure as a chest x-ray, which is about the same amount natural background radiation that most people get in 10 days of normal living. However, a CT scan exposes you to about 8 years of background radiation exposure! (27) Hence, we generally DO NOT recommend CT scans to diagnose CCI.

MRI Imaging Pitfalls

Before we get into the topic of imaging and MRIs, it's critical to point out that a routine cervical MRI has limitations in CCI patients. Let's explore that a bit.

An MRI (Magnetic Resonance Imaging) machine is a big magnet. It applies a huge magnetic field which aligns the small molecules in your body and then removes that magnetic field and lets these flip back to their normal state. When that happens, these molecules produce a radio signal that is reconstructed by the computer into an image.

The Good News

Static measurements like Grabb-Oakes and the CXA can be measured on most routine cervical MRIs. This may help make the diagnosis of CCI. The problem is that, as you'll see below, movements like flexing your neck forward and backward usually enhance the information gathered. That kind of motion can only be picked up on a specialized "Stand-up" MRI unit and not on the standard "lie face up in a tube" type. More on that below.

The Bad News

What an MRI can detect depends on the specific coil used. This is something placed around the body part that acts as an antenna to pick up the signals coming out of your body that are generated by the big magnet. There is a different coil used for each body part.

The usual cervical MRI only images from C2-T1 because that's what the neck coil is optimized to image. However, the pathology in a CCI patient is above C2. In fact, to get good MRI pictures above C2, a head coil is often needed. Hence, having your doctor write a script for a routine cervical MRI may not be helpful in getting to a diagnosis.

Imaging Resolution and Strength

It's also critical to understand that imaging strength is directly related to the resolution or quality of an image. Hence, whatever type of static MRI you get, you should make sure that you get the best possible

image. In addition, getting imaged on a more or less powerful MRI machine is agnostic to your insurance benefits. Meaning, getting imaged on a 2-million-dollar 3.0 Tesla MRI is covered by insurance just as much as a getting imaged on an old-school 0.3T open MRI that someone would pay you to haul away for parts.

MRI imaging strength is measured by a unit called a Tesla (T). That's for early 20th century electrical engineering genius Nicolai Tesla and NOT the car company. One Tesla is the magnetic field strength of the earth. Hence, 0.3T means about 1/3 that amount.

Here is how they rate out:

- Worst quality images-0.3-0.6T
- Good quality images-1.5T
- Best quality images 3.0T

You can find this out by asking the center the "field strength" of the machine. They should report it as above.

Realize that any open or Stand-up MRI has a lower field strength than most high-quality lying down MRIs. This is a regrettable trade off. For example, in order to be able to image someone in a machine that allows for motion, you need more room, and hence the magnetic field will have lower strength to make that work. However, more information about an instability condition like CCI can be gathered when patients move. Hence while field strength may be important in a static MRI, we're never going to see a Stand-up MRI with 3.0T.

The "Best" Type of Imaging for CCI?

In our clinic, we like using DMX (discussed below), which allows for movement that approximates reality. Our second choice would be movement-based MRI or a rotational CT scan. Static MRI of the upper neck can sometimes be helpful and there are times that we can see abnormal measurements on a routine neck MRI.

However, this preference for DMX is not universal. For example, many neurosurgeons prefer routine static MRIs where some of the measurements shown below can be measured.

A Gold Standard?

While it's always great if any diagnosis has a gold standard test to determine if you have or don't have the disease, that doesn't exist in CCI. Hence, different physicians all use different tests to get to a diagnosis. While that makes it harder for patients, who understandably want certainty, it's reality.

Types of Imaging

Imaging can be broken down into two main categories: static and dynamic. Static means that the patient doesn't move during the imaging. This can be a problem for diagnosing instability as that, by definition, involves movement. To solve that issue, more recently applied imaging techniques use movement, and this is called dynamic imaging.

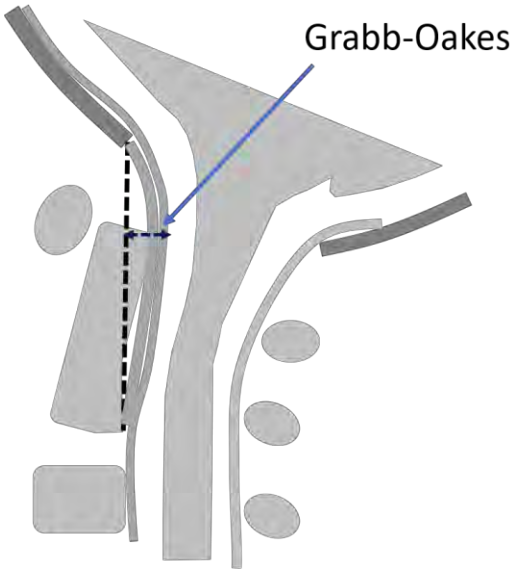
Before I jump into explanations of all of the different types of measurements and imaging used, realize that this chapter is yet another example of "threading the needle" between a book that can be read by

patients, but that has enough detail to be handed to a physician that needs to be educated. Hence, it's OK if you jump through parts of this chapter that are in too much detail for you.

Static Measurements

Grabb-Oakes

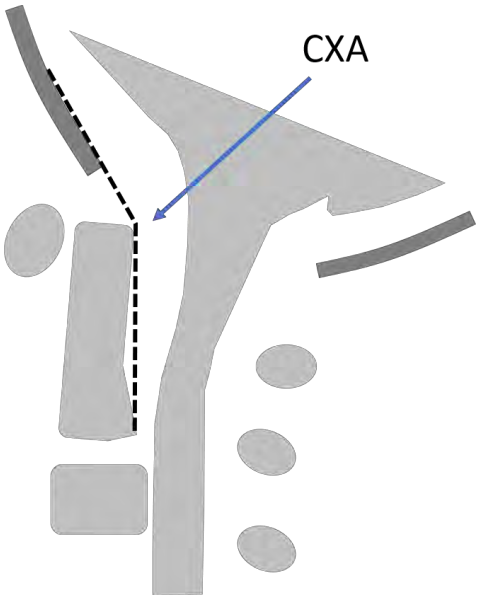
This is the distance between a line drawn from the front of the spinal canal (basion) and the back inferior corner of the C2 bone and the back of the dens. Given that the C2 bone (axis) is held against the C1 bone (atlas) by the transverse ligament, a high Grabb-Oakes measurement can be due to a lax transverse ligament or a large pannus. An abnormal measurement is often considered to be 9 mm or more (2,5). With the advent of flexion-extension MRI, it's easier to measure the G-O measurement when the patient moves, which provides more information. See this video for more on the G-O measurement [https://youtu.be/4f-Yi9fuKD0]. However, a new upright MRI measurement study calls into question the utility of using flexion-extension differences due to the large overlap in the range of values between those two movements (28).



CXA (Clivo-Axial Angle)

This is the angle between the clivus (the inside front area on the bottom of the skull) and the back of the dens (C2). The problem being measured here is the skull is falling forward on the upper neck which can cause irritation of the front of the brainstem and upper spinal cord. This movement is controlled by strong ligaments in the back of the neck such as the nuchal, supraspinous, and interspinous ligaments. It's also controlled by the posterior atlantoaxial membrane (PAOM, a ligament at the back of the spinal canal) and to a lesser extent the transverse ligament. The tectorial membrane is also involved. Abnormal is a bit different when reported by different authors. Less than 150 degrees (4) was originally reported with others stating that normal is between 145° to 160° in the neutral position (3). More recent research has shown that CXAs approaching 120 degrees are likely pathologic, causing too much stretch on the spinal cord and brainstem (27). The differences in opinion are likely due to the fact that things like kyphosis (forward head posture) and male/female sex can change the angle. I tend to use a cut off of 125 on an upright MRI. Patients in these lower ranges of CXA with obvious pressure on the neural structures

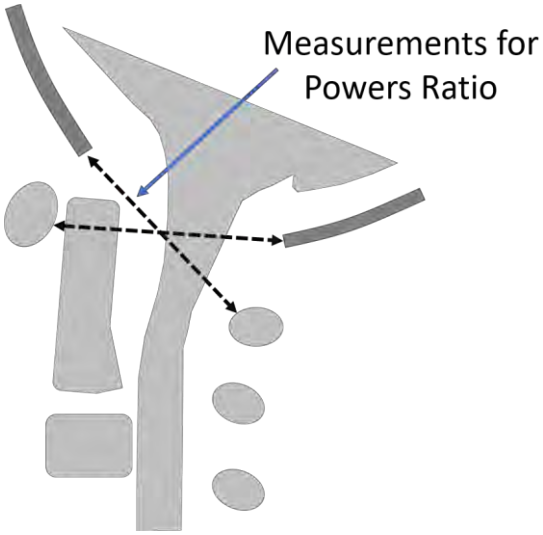
should also consider getting an advanced Diffusion Tensor Image which can show if there is damage in the spinal cord/brainstem.



Power's Ratio

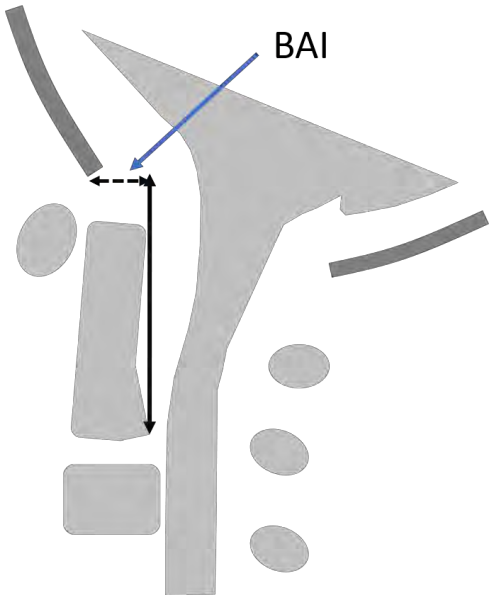
This is another measurement that determines if the head has moved forward on the upper neck. This one is more complex, requiring two lines (one between the basion and the back of the spinal canal at C1 and another between the opisthion and the posterior aspect the front of the atlas) and then some division is applied. If the calculated measurement is less than 1, the ratio is normal, if it's >1 then it's abnormal (2). The Powers Ratio measures whether the head is aligned properly on the upper neck bones. For example, if the number is greater than 1, the head is too far forward on the spine and multiple ligaments might be injured.

See my video here for more information [https://youtu.be/mQw7Sx5QA2c].



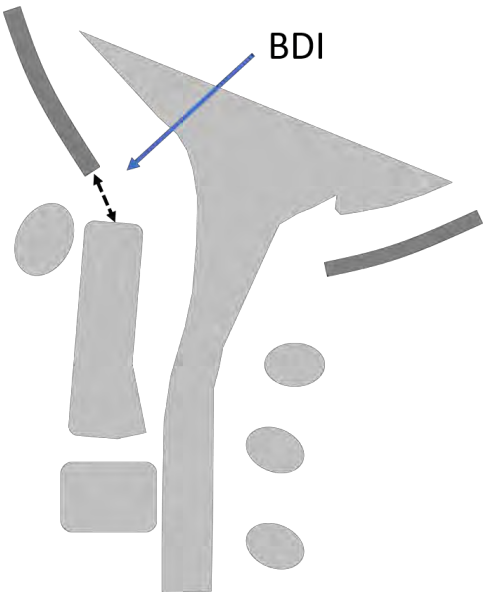
BAI (Basion-Axial Interval)

This is the distance between the basion (front of the skull) to the back of the dens of C2 (axis). The problem here is that CT scan studies have shown the usual normal of 12mm to be unreliable with normal ranging from 9-26 mm. This can also be measured in flexion and extension where >4mm difference between the two positions is considered abnormal enough to warrant surgery. The ligament most responsible for keeping this measurement in check is the transverse ligament. A recent study on upright MRI patients with neck pain but not CCI showed that the normal in these 50 patients could be as low as less than 6.2 mm. This study also showed that 95% of patients had less than 1mm difference between flexion and extension.



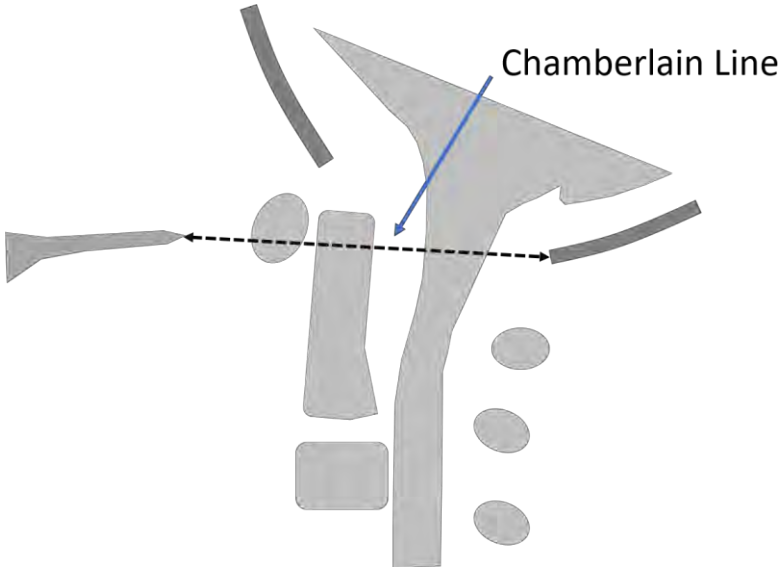
BDI (Basion-Dens interval)

This measurement focuses on vertical instability. This would be damage to the ligaments preventing vertical translation of the head like the PAOM, tectorial membrane, AAOM, apical, and SAAOL. The normal value is less than 12 mm on x-rays and 8.5 mm on CT scan (2). Like the BAI, it can also be measured in flexion and extension, but newer research suggests that this measurement on an upright MRI may not be meaningful (28). The BDI change can also be measured as traction is applied to the head. The most recent upright MRI study on this topic suggests a lower cut off for abnormal on upright MRI should be around 9mm allowing for a 5% false positive rate (28).



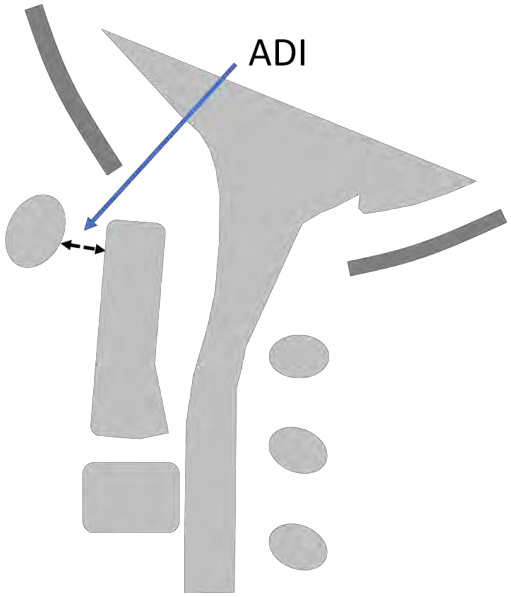
Chamberlain Line

This is a line drawn from the back of the hard palate of the skull to the back of the hole in the skull (opisthion). If the dens of C2 is more than 3mm above this line, this indicates cranial settling or that the skull is too low on the cervical spine. This is also called basilar invagination. Both of these conditions are more extreme than are usually seen in most CCI patients. Meaning in in purest neurosurgical sense, both cranial settling and basilar invagination have the dens touching the brain stem.



ADI (Atlantodental Interval)

This is the distance between the front of the C1 (atlas) and the dens of C2. Normal has been considered to be 2-3mm (2). If this is greater than 1-2 mm in older patients, the transverse ligament may be injured. Like many of these measurements, the ADI can be measured in flexion as well to see if it increases, as that stresses the transverse ligament. In younger patients, there is more natural distance due to cartilage between the dens and the atlas, hence the cutoff for abnormality increases to 2-3mm (36).



Upper Cervical MRI

Note that a routine neck MRI rarely shows these upper cervical structures. Much of that has to do with the wrong type of coil used to collect the information which is designed to an image from the neck and not the upper neck/head. This means that ligaments like the alar, transverse, and others just don't show up on the usual cervical MRI.

However, if a specialized MRI is taken, the upper neck ligaments can be imaged. This requires a head coil and other specialized image sequences. Once we have those images, you can look at how the ligaments appear and grade them based on a scale published by Krakenes (6). While we look at this information, the problem is that it's not showing whether those ligaments work functionally to stabilize the head on the neck. That would require movement-based imaging. If you want to learn more, see my video [https://youtu.be/ZQaztACMzbk].

CSF Flow Imaging

One of the more promising new technologies out there is using a specialized MRI to measure the flow of cerebral spinal fluid though the upper neck. This is critical because many problems that surround CCI can restrict the normal flow of the brain's fluid through this area. The world's expert in this area is Scott Rosa, DC of upstate New York. He's been using this technology in CCI patients for years and can show improvements of CSF flow through the upper neck before and after specialized chiropractic adjustments (see http://rosaclinic.com/)

Dynamic Imaging

Flexion-Extension X-rays

These are x-rays where the patient is asked to look down and then an image is taken and then they are asked to bend their neck back and look up and another picture is taken. The goal is to see if the neck vertebrae move too much against one another, which they can only do if the ligaments are damaged. For CCI, the one measurement that can be looked at here, is the ADI when the neck is in flexion. In addition, many CCI patients have also damaged other neck ligaments in the lower neck that can be evaluated here.

One of the biggest issues with this type of imaging is that many radiology techs don't get enough movement out of the patient to allow proper measurements to be made. For these films to be diagnostic, the patient needs to flex and extend as far as possible, despite the instructions from the tech taking the picture. Rad techs often tell people to move minimally because it's easier for the tech, but make sure you're a rebel here and get as much motion as possible!

Upright MRI

MRI of the neck is usually performed with the patient lying face up in a tube (supine). Upright MRI allows a patient to be imaged in a weight bearing position. This places more natural stress on the upper neck and other ligaments, so it's possible that measurements that weren't indicative of CCI lying face up in the usual MRI scanner could be become positive when imaging the patient this way. Having said that, normal values obtained while imaging people supine may not apply to upright MRI.

Upright MRI also allows for motion. Hence, the patient can do things like bend or extend the neck. This can give valuable information about whether the ligaments perform normally or allow too much motion. To learn more, see my video here [https://youtu.be/rpHv0AalJ4Y].

Functional CT (C1-C2 Rotatory CT)

This is a specialized CT scan where the patient rotates their head and the movement between the skull, C1, and C2 is measured (7). Instability is indicated by rotation of the skull on the atlas of more than 7 degrees, and of the C1/C2 joint of more than 54 degrees.

DMX (Digital Motion X-ray)

DMX uses a fluoroscope which is real time x-ray imaging and captures video of how the upper neck bones move. The technology is the only one that allows for natural movement of the patient as all others are contrived and artificial in some way. DMX allows a patient to move in one direction at various velocities and accelerations. The patient then must quickly stop and go another direction. This is very different than a neck flexion-extension x-ray where the patient only moves in one direction maximally and then a picture is taken.

The measurements that are important for CCI patients here are C1-C2 overhang and ADI. C1-C2 overhang means that the patient bends their head to the side and the doctor looks to see if C1 slips sideways against C2. If that happens beyond 3mm with also a difference in the distance between the dens and the atlas, then the alar ligament on the same side may be injured. The ADI can also be measured. To learn more, see my videos [https://youtu.be/nPBW4Bk-8l4 and https://youtu.be/6MRnZw2BpV8].

A Word of Caution About DMX Reports!

My job as your physician is to determine if your imaging findings are normal and expected for your age and part of normal degenerative changes or more likely than not causing your symptoms. With measurements like those discussed above (overhang and ADI), we know which levels of extra movement are more likely to cause symptoms. For other things commonly reported on DMX, these are usually normal degenerative wear and tear and NOT "damage" as often reported in written DMX reports.

The things we look for on DMX that are important in the diagnosis of CCI include: C1-C2 overhang more than 3-4mm, an ADI in flexion of more than 2-3mm in older people and 4mm in younger people, and C2-

C3 translation in either flexion or extension of more than 1 mm. Findings that are not as important include: facet gaping, lower neck translation under 3.5mm, spinous process rotational measurements, and the skull hitting the atlas on extension.

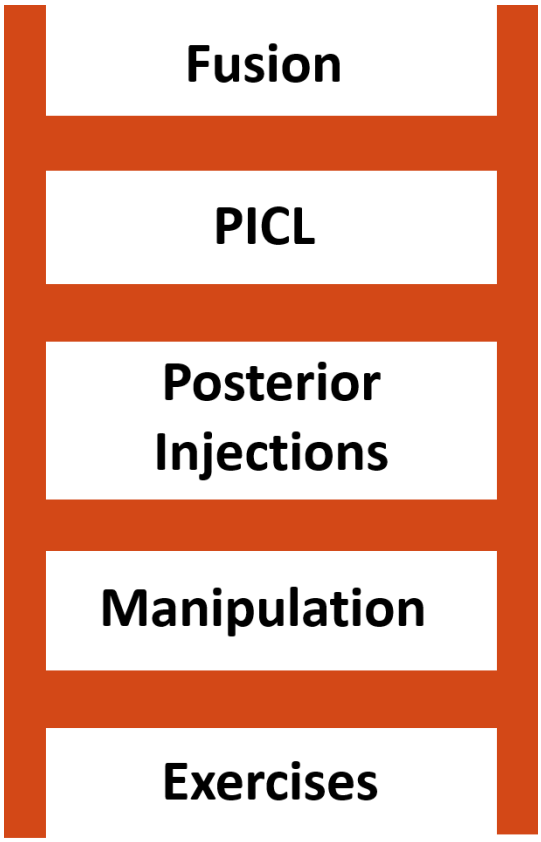
This is also a good point to mention that we generally only treat symptomatic areas. That means that many of these degenerative areas, if they are asymptomatic (which is likely) won't get treated during a PICL procedure.

Chapter 5-Treatment

Understanding the Ladder of Invasiveness

How do physicians look at medical care for their own families? They use "the ladder". What's that?

If you want to keep patients as safe as possible, you only expose them to the least risk needed to get the job done. For CCI, that system creates a ladder that looks like this:



You'll notice at the bottom I have listed exercises. Things like physical therapy and alternative healing techniques like craniosacral would be in this category.

Next up would be manipulation of the upper neck. These are things like NUCCA or AO chiropractic. These are slightly more invasive than physical therapy.

Then we get into posterior injections (from the back), which could be prolotherapy of the ligaments in the back, facet injections, epidurals, etc. This is more invasive than manipulation.

Next up are anterior (from the front) injections which include the PICL procedure. This means injecting the deep ligaments that hold the head on. Since this is a newer procedure where fewer have been done than posterior injections, it's automatically considered higher risk than more commonly performed posterior injections.

Finally, at the top of the list are spine surgeries like detethering, decompression, and fusion. For example, a surgical detethering procedure would be more invasive than PICL since it involves open surgery and destroys a part of the spine. Even more invasive would be fusion which is where screws, rods, or plates are placed to make sure parts of the spine or a joint can never move again.

So how can you use the ladder like I would to protect my wife and kids? Anything lower on the ladder that applies to that injury or problem that may work needs to be tried first before moving up the ladder. That way you don't get exposed to more risk than you need.

Conservative Care

Conservative care for CCI breaks into a couple of common areas:

- Bracing
- Physical therapy
- Chiropractic
- Curve restoration
- Cranio-sacral

Bracing

Neck bracing is an interesting topic in the CCI community. On the one hand, you have patients that find the right neck brace and swear by it. On the other hand, you have patients who can't find a brace that will keep their head/neck in the position where it's happy with less pain. For those who like braces, this is a two-edged sword. Why? Because stability requires normal ligaments and muscles. Hence, using a neck brace for an extended period of time can make the neck muscles weaker, which can make the instability worse. So, if you use a brace, use it only for short periods. For example, some patients will only use one when travelling or exercising. Some need it more often or they can't function. Just remember, like all CCI treatments there are advantages and disadvantages.

Physical Therapy

One of the things that's almost diagnostic for CCI is that patients do poorly with active physical therapy that focuses on strengthening. This response can change if the patient has had their upper neck stabilized through the PICL or a fusion procedure. At that point, especially with the first two treatments, exercises are encouraged to slowly help the patient build back the upper neck muscles that stabilize the head on the neck. Of note, if the patient is fused, normally some muscles are destroyed by the procedure. More on this later.

However, there are physical therapists with specialized manual knowledge who can help CCI patients. Meaning that these therapists have years of additional training, so they represent less than 1 in 100 of all PTs. A helpful guide to finding an upper cervical expert is finding a therapist who knows the upper neck. Here are some resources:

- IPA Physical Therapy [https://instituteofphysicalart.com/]
- Ola Gimsby Manual Therapy [https://www.olagrimsby.com/]

Other types of PT to consider that don't focus on the upper neck, but do focus extensively on postural correction that may help the upper neck include:

- Feldernkrais [https://feldenkrais.com/]
- Egoscue [https://www.egoscue.com/]

Chiropractic

While physical therapists have few experts who know how to treat this specialized area of the body, that's not true for chiropractors. In particular, there is NUCCA [https://nucca.org/], which is a national upper cervical chiropractic association that qualifies and trains these providers. Many of my patients swear by NUCCA chiropractors who are able to help their symptoms. Usually, if they find their way to me,

they will find one who can get temporary relief from hours to days to weeks, but then things come out of place. On the other hand, for other patients, NUCCA is not helpful.

Another type of upper neck chiropractic is Atlas Orthogonal (AO). This is very similar to NUCCA, but instead of a hands-on manual adjustment, a specific low force instrument is used to precisely move the atlas back into place.

How do both of these differ from traditional chiropractic? Many chiropractors will perform "long lever arm" neck manipulations where they suddenly twist the skull. NUCCA instead uses a precise, noninvasive spinal adjusting technique. This uses precise and objective x-ray views of the head and neck, mathematical measurement and analysis to plan the specific low force adjustments used by these providers.

Curve Restoration

Your neck is born with a natural shallow c-shaped curve. This distributes the forces from your head to the discs in the front of the neck and the facet joints in the back. In our modern society, looking down at computers and phones or neck trauma can cause the neck to straighten or develop a reverse curve. That puts too much force on the discs in front and can cause too much force on the upper neck ligaments as well. Hence, we've seen several CCI patients get improvements with curve restoration. There is an online credentialing organization where providers can be found [www.idealspine.com]

Curve restoration involves forward traction to increase the curve by pulling the neck forward (not to be confused with axial traction that pulls your head upwards). Specialized machines are used to do this and at home devices are also available. It should be noted that only some CCI patients find this helpful (usually fewer than find NUCCA or AO to be helpful).

Injections

This is a big topic. Before I get too far into it, it's good to spend a few minutes on how different injection procedures stack based on complexity:

Level 5	Image guided injections that are extremely rare. PICL is an example, done with fluoroscopy and other modalities like endoscopy. Only a handful of providers worldwide can offer.
Level 4	Highly advanced image guided injections into things like upper neck facet joints (C0-C2), done with fluoroscopy, take many years to master. Only 0.1% of providers offering spinal injections.
Level 3	Advanced image guided injections into things like lumbar and cervical discs, done with fluoroscopy, take months to years to master. Only 10% of providers offering spinal injections.
Level 2	Image guided injections into things like lower neck facet joints or epidural. Often done with fluoroscopy, take months to years to master. Only 20% of providers offering spinal injections.
Level 1	Simple muscle trigger point or posterior spinal ligament injections (prolotherapy). Often done blind, take hours or days to learn. Many providers can do these safely.

Why? We often see patients who get confused that Level 1 injections as shown have something to do with Level 5 injections. For example, blindly injecting prolotherapy solution into the back of the neck has little to do with using endoscopy and fluoroscopy to inject stem cells into the alar and transverse ligaments (PICL). To use another analogy, the skill level needed to do the level 1 injection can be taught

in a few hours and the skill level needed to perform a level 5 procedure would require many years to master.

For CCI patients, the different skill levels break down as follows:

Level 1-Posterior prolotherapy. This is injecting the easy to reach ligaments usually with no imaging guidance (blind or palpation guided). Sometimes the doctor will use a fluoroscope, but not in the same way a more experienced and highly trained doctor performing higher level injections would use that machine. The Level 1 doctor is just using that fluoroscope to make sure they have hit certain bone landmarks, whereas the higher-level physician with more training is using that machine to make sure that they are in certain joints by injecting radiographic contract. I could teach the first technique in a few hours, but the second takes months to years to master.

This is not to say that in the right patients with general neck instability, posterior injections are worthless. They can be very helpful. However, the vast majority of CCI patients that try posterior injections don't get relief and functional benefit. **This is a concern as MANY CCI patients spend thousands to tens of thousands of dollars on posterior injections with prolotherapy, platelet-rich plasma, or bone marrow stem cells that ultimately don't work. This is despite the promises of a few practitioners that these procedures will be highly effective.**

Why won't posterior injections help most CCI patients? Because the ligaments that need the most attention can't be reached via this approach.

Level 2-Lower neck facet or epidural injections. This is using x-ray or ultrasound guidance to inject the C2-C7 facets or spinal nerve levels. These procedures can usually be performed by any competent interventional pain physician.

Level 3-This is a bit more intense and for CCI patients, this might include injecting the C2-C7 neck discs if needed.

Level 4-For CCI patients, a major issue is finding a physician who has the rare experience of injecting the upper neck facet joints (C0-C2). Why? These are much more technically demanding and thus more rarely performed.

There are only a handful of US physicians who have injected these joints more than a few dozen times. Since repetition in medicine breeds competency, if you've only ever done something a dozen or two times, you're not an expert. Hence, these are physicians who have injected these joints at least 100 times or more. For comparison, our clinic physicians have injected them much more than 1,000 times. **As a result, we do not recommend any clinic outside of our Colorado site to perform these level 4 injections.**

Level 5-Finally, now we're at the level of a physician who would be capable of performing the PICL procedure which will be discussed later. This would only be a handful of experienced interventional spine physicians who would also be willing to take the time to learn how to treat this patient population. At the time of this writing, the only site that is qualified to perform this procedure is the Centeno-Schultz clinic in Colorado.

I can't stress enough that the type of injections you need depends on the type of CCI diagnosed! For example, while a type 3a CCI can be treated with posterior ligaments injections (levels 1-3 above), a type 2b CCI requires direct injection of the alar ligaments which is a level 5 PICL procedure.

Which type of injections can treat which kinds of CCI? Type 3a can be treated with posterior injections. Type 1a may also be treated with posterior injections, but the PICL may also be required. For types 1b, 1c, 1, d, 1e, 2a, 2b, and 3b, the PICL procedure is usually required.

A Note on Umbilical Cord "Stem Cells"

In the United States, we now have many practitioners offering level 1 injections with Umbilical Cord "stem cells". Our lab has tested many of these Wharton's Jelly products and our research team and that of CSU Translational Medicine Institute clearly show that these products have no living or functional mesenchymal stem cells (35). Hence, any clinic offering this treatment as "stem cell" is a scam.

Chapter 6-Is There Another Way? PICL

What if, instead of fusing the upper neck with hardware, we could instead prompt the damaged and loose ligaments to heal? That's the concept behind the PICL procedure. This acronym stands for **P**ercutaneous **I**mplantation of the **C**raniocervical **L**igaments.

Who Is a Candidate for this Procedure?

Patients with CCI who continue to have disability despite conservative care and who have been qualified as discussed in earlier chapters. We tend to use a DMX study the most often to nail the CCI diagnosis and less often, a movement-based MRI.

Procedure Summary

This is a procedure where we use endoscopy and fluoroscopy to precisely guide a needle into the craniocervical ligaments from the front. We start by imaging the back of the throat using endoscopy and preparing a sterile field. A special 3D printed mouthpiece is also used to keep the mouth open and the tongue depressed. A needle is then placed on either side of the uvula and fluoroscopy is then used to

guide the needle into the craniocervical ligaments. Bone marrow concentrate is injected to help ligaments heal.

What's Injected?

We use bone marrow concentrate prepared in a cGMP class clean room. This means that we start the procedure with a bone marrow aspiration. Most patients want to be put asleep with an IV for this part with some just getting local numbing. The bone marrow aspirate is then processed in our lab to concentrate the stem cell fractions.

Two Types of PICL Procedure

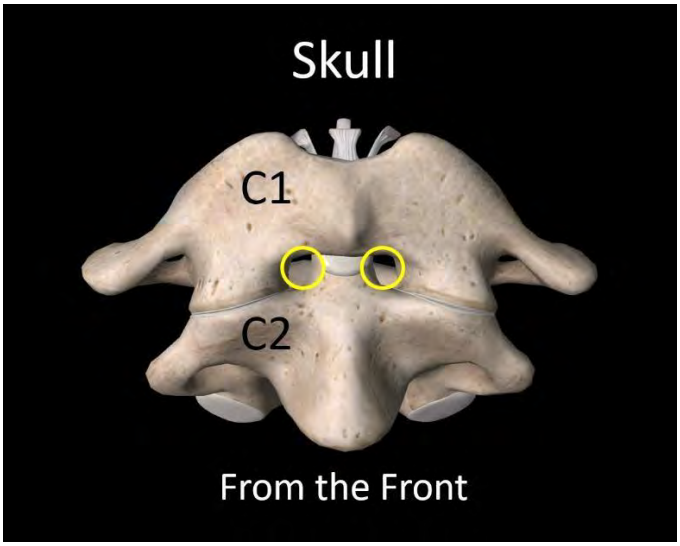
There are two types of PICL procedure: above the atlas and below the atlas.

Under the Atlas

This approach allows the doctor to access the transverse ligament, the accessory and lower alar ligament, tectorial membrane, and the anterior longitudinal ligament at C2-C3.

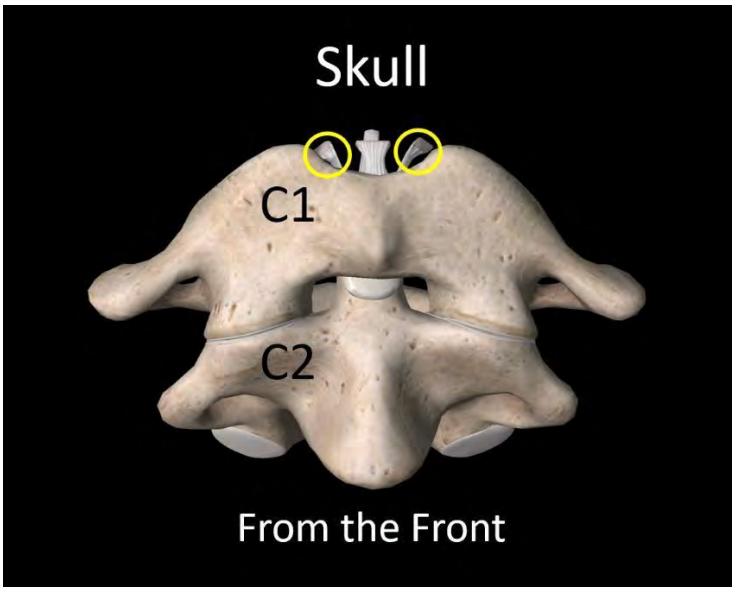
Above the Atlas

This approach provides access to the upper alar ligament, transverse occipital ligament, anterior C0-C1 facet capsules, sAAOL, AAOM, and tectorial membrane.



There is a small hiatus (opening) between the C1 and C2 bones where a needle can be threaded (yellow circles). Once through that area, the lower alar, accessory, transverse, and other ligaments can be reached and injected.

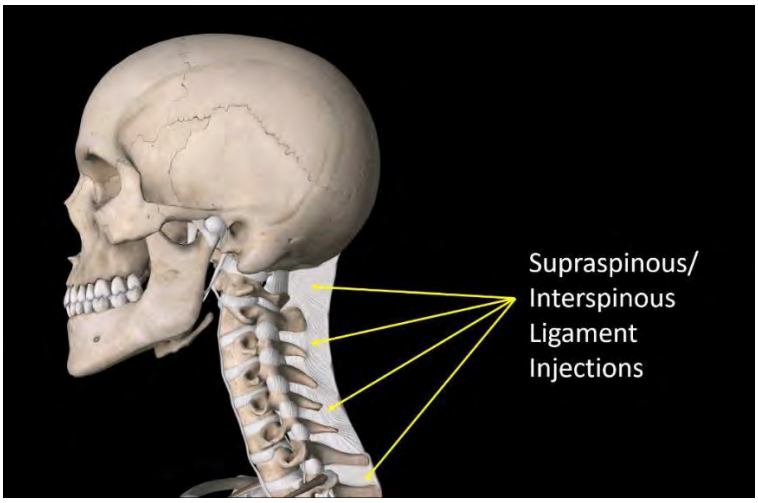
Over the Atlas



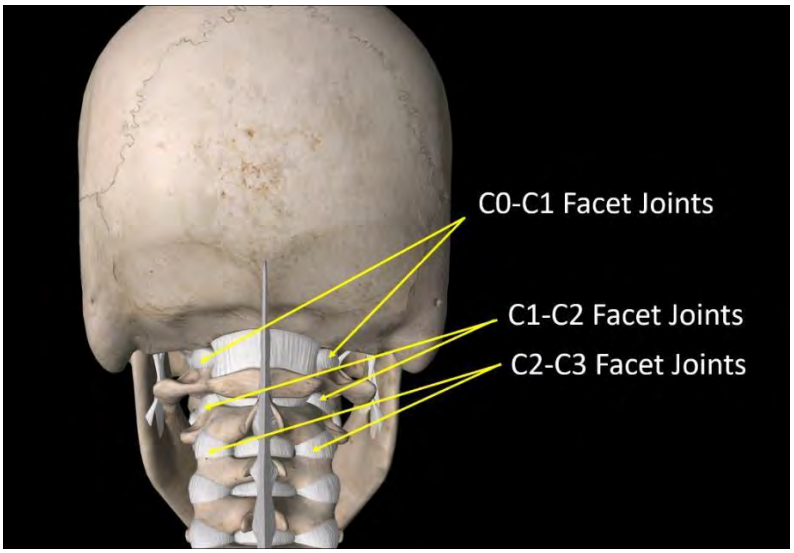
The needle can also be directed above the C1 bone to reach the upper part of the alar ligament and the ligaments that extend upwards from the atlas to the skull (SAAOL, AAOM, apical, cruciate).

We begin almost all PICL procedures with a below atlas injection and then determine how much coverage of the transverse, accessory, and alar ligaments was obtained. If the patient is sensitive to procedures and is centrally sensitized, to avoid a big post-procedure flare-up we will usually stop there if coverage is adequate. If the patient isn't centrally sensitized, then we go above the atlas as well during the first procedure. Any patient who didn't get the above atlas on the first procedure, assuming a normal post-injection recovery, will get it on the second procedure. In addition, in almost all patients, if inadequate ligament coverage is detected from the below atlas injection position, then we will add the above atlas procedure automatically.

Posterior Injections



Nearly all PICL patients also receive posterior injections with bone marrow concentrate as well. This will be at least the supraspinous, interspinous, nuchal, vertebrodural, and PAOM ligaments.



Many also get upper cervical facet injections using sophisticated fluoroscopic guidance. These are often the C0-C3 facets as shown above. Some patients also need occipital nerve hydrodissection. This is where the growth factors from the patient's own platelets are injected around the injured nerves at the back of the skull to help them heal.

What to Expect

The Exam

The patient is first seen for a hands-on evaluation in the office. The purpose is largely to define other areas outside of the PICL that need to be treated. For example, the doctor will try to answer questions like:

- Localizing which upper neck facet joints are largely involved
- Which occipital nerves may be causing headaches?
- Whether the superficial cervical plexus under the SCM has issues?
- Is the TMJ involved?

In addition, many CCI patients have developed other problems in various body areas. For example, the shoulder, upper back, numbness or tingling in the arms or legs, pinched spinal nerves, SI joint instability, etc.… The goal is to see which of these issues are likely to resolve once improved upper neck stability is achieved and which problems have taken on a life of their own and need separate treatment.

The Bone Marrow Aspiration

A Bone Marrow Aspiration (BMA) is a procedure where the doctor takes the liquid portion of the bone marrow from the back of the pelvis. The goal is to tap the stem cell rich bone marrow aspirate. Before we proceed, we'll review some discussion on the best way to improve your stem cell counts.

While nobody is yet 100% sure based on high-quality research, we've seen a few things improve stem cell counts:

- Intermittent fasting prior to a procedure. One good resource to investigate is the Prolon system (also called a fasting mimicking diet)-see https://prolonfmd.com/

- The Stem Cell Support Formula offered by Regenexx (see https://store.regenexx.com/)
- Reduce your chronic inflammation-Supplements like fish oil and turmeric can be helpful here, see https://regenexx.com/blog/running-on-empty-fish-oil-and-my-ankle/mat

An IV is started and the patient is taken back to a procedure room, here the doctor will usually use IV sedation. Hence, it's critical that the patient not eat anything for 6 hours before or drink anything for 2 hours before the procedure. The doctor will get the patient sleepy or asleep and then numb the back of the pelvis area. A specialized needle called a trocar is used to access the bone near the PSIS (dimples of Venus). Bone marrow aspirate is drawn using ultrasound or x-ray guidance and then sent via sterile transport to the lab. The area is bandaged up and the patient is sent to recovery. This part usually takes 30-40 minutes. Sometimes additional blood will be drawn which will be collected from a vein at this time. After this is done, this is a good time to hydrate before the PICL procedure, but please don't eat.

The patient is then free to go home, hang out near the office, or head back to their hotel until the procedure in the afternoon.

The Processing

Because the PICL is a hyper sterile procedure and to avoid infection in this area, we use a full cGMP class cleanroom to process cells. This means that this is a specialized space with HEPA filtration and strict entry and exit criteria.

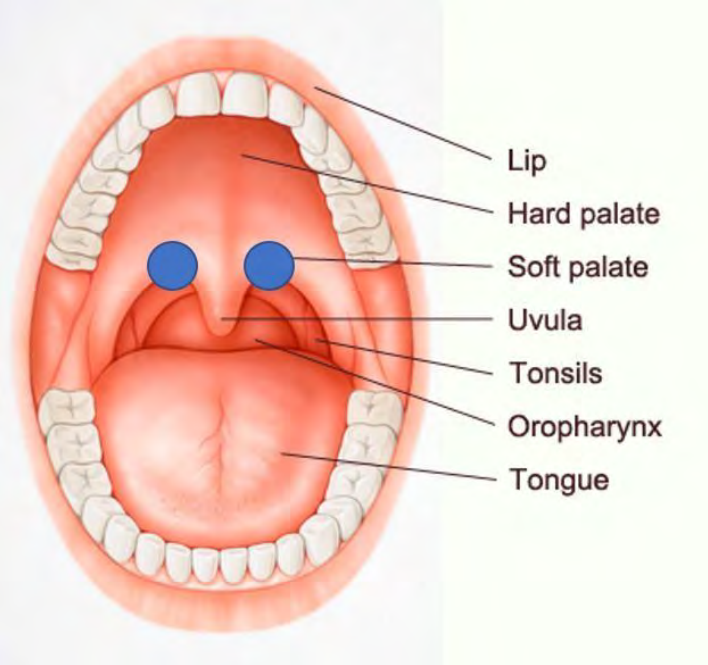
You may never see this facility when you're in the office, but you can ask for a tour of it. It's a 1,200 square foot specialized room on the first floor of the practice (the clinic is on the second floor of the building). This is where our processing staff works on one patient sample at a time in an ISO 5 biologic safety cabinet with laminar flow. The bone marrow is centrifuged and then the two stem cell fractions are combined for later injection. Platelet-rich plasma, platelet lysate, and platelet poor plasma are also made either from the bone marrow or peripheral blood. All of these are placed in sterile packaging which is labelled with the patient's name and date of birth and sent back up to the clinic where it's stored in a specialized 4C medical grade refrigerator until the patient's procedure.

The PICL Procedure

The patient returns in the afternoon and is first checked into a patient room. An IV is started and then the patient meets with the nurse anesthetist. Some initial relaxation medications are given and then the patient must perform a sterile chlorhexidine mouthwash. They are then taken back to the procedure room where more medications are given once the patient is placed face up on the table.

The anesthetist then matches the right 3D printed mouthpiece for the patient. We have a number of different designs we've designed and used. If this feels uncomfortable, just realize that more medication can be given IV to help with the anxiety of this part of the procedure.

Next, the procedure room staff then clean off the front of the mouth area with iodine and drape the patient as they are getting very sleepy. The doctor usually comes in around this time and then is dressed by the staff in a sterile gown, gloves, mask, and hat.



The doctor then uses endoscopy to visualize the back of the throat while using sterile cotton swabs to apply iodine several times to define a sterile field. The doctor will also inject a numbing medication at the back of the throat and then clean again. The injection needles are then placed on either side of the uvala (blue dots above) using an endoscope and c-arm fluoroscopy and the procedure begins. Usually, the patient doesn't remember much after the drapes go on.

For the PICL procedure, anesthesia keeps the patient fully asleep using multiple different medications. These include versed, fentanyl, propofol, and sometimes ketamine.

After the PICL procedure is complete, the staff will wake the patient up partially so that they can help get turned face down. The sterile prep will be repeated, and the patient will be given more medications. The doctor will complete the posterior injections and then the patient will be discharged to the recovery area.

In Recovery/PACU

The patient will wake up in recovery. It's very common to have pain at the back of the throat and back of the head. If the patient has a history of nausea, then this can flare up. Same with other common symptoms like dizziness, imbalance, etc.… In addition, a drug called ketamine may be used during the anesthesia, so you may want to inform loved ones that the "ketamine stare" may be present and that you'll be a bit "off" for a few hours. The patient typically spends 1-2 hours in recovery.

In the Hotel or At Home the First Night

The patient is typically in pain and can have all the symptoms described above. Pain medications and antibiotics and possibly anti-nausea medications will be provided via script. It's best to take pain medications about an hour after returning back to the hotel or home. Realize that we want you to stay ahead of your pain, so don't be shy about taking the prescribed pain medication amounts.

It is very common to be **very** flared up after the procedure. Hence, this isn't a cause for panic. Here are some things that indicate that you may need to call your doctor:

- Disorientation or confusion that isn't related to the anesthesia or medications
- Fever, chills, sweats
- New neurologic symptoms that you have never experienced before

Post-Procedure

About half of the patients have a "honeymoon" period where they may be in pain, but they feel stable. Hence, they feel better for the first 1-2 weeks because the swelling causes the loose ligaments to tighten. Then this goes away as the procedure caused swelling reduces. Some sense of stability and improvement in symptoms returns once the patient is able to lay down new tissue promoted by the stem cells in the bone marrow concentrate.

The other half of the patients experience a flare-up with no honeymoon period. They generally have increased symptoms for 1-3 weeks and then things calm back down to baseline. A handful of patients can experience longer flare-up periods lasting 1-2 months. An even smaller number of patients may have flare-ups lasting a few months.

We generally look at months 3 or 4 to see if there has been any benefit from the procedure. Improvements by or after month 4 are the most common. A few patients might get benefits later than that time period, but this is more the exception rather than the rule.

Understanding How Ligaments Heal

To get a sense of the phases that patients will cycle through after the PICL procedure, it's important to look at how our ligaments heal after a regenerative injection. The first phase is inflammation, which lasts for 1-2 weeks after the procedure. This is when the body calls in the right cells to initiate a healing cycle. The next phase is remodeling, when the body is laying down new tissue. This phase can last for 2-8 weeks. Finally, the third phase is maturation, when the newly laid down collagen fibers in the ligament align to allow the ligament to gain strength. This begins 2 months out, with a shorter final healing period of 3-4 months in younger patients and a longer period of up to 6-9 months in older patients.

The Timing of PICL Procedures

Given that it takes 3-4 months to see results, we usually sequence these procedures every 4-6 months. Patients who want to wait a full 6 months after the first procedure will get to that place after the flareup where they can assess if the procedure helped. Some patients who want to compress the time frame of the whole process by getting procedures done every 3-4 months in rapid fire could stay flared up for most of the time, hence this if the patient finds themselves flared up for a longer period of time than a few weeks, it's generally recommended that they consider waiting until the procedure flare-up ends before scheduling the next procedure.

On the other end of that spectrum, waiting a year or more between these procedures could be counterproductive. Meaning that the procedures work by building new tissue. If the rate at which new tissue is built is exceeded by its breakdown through daily wear and tear because of instability, then the patient may feel better for a while and then backslide.

The average number of procedures required is 2-4 with some patients being “one and done” and some going onto five procedures.

What to Expect-Two Steps Forward and One Back

For some patients, their recovery will be linear, meaning after the flare-up, each procedure will provide benefit until they eventually achieve maximum healing. For others, it may be two steps forward and one back. Meaning there may be ups and downs in their recovery journey. For still others, this type of procedure may not be effective, and they may need to consider fusion.

Results?

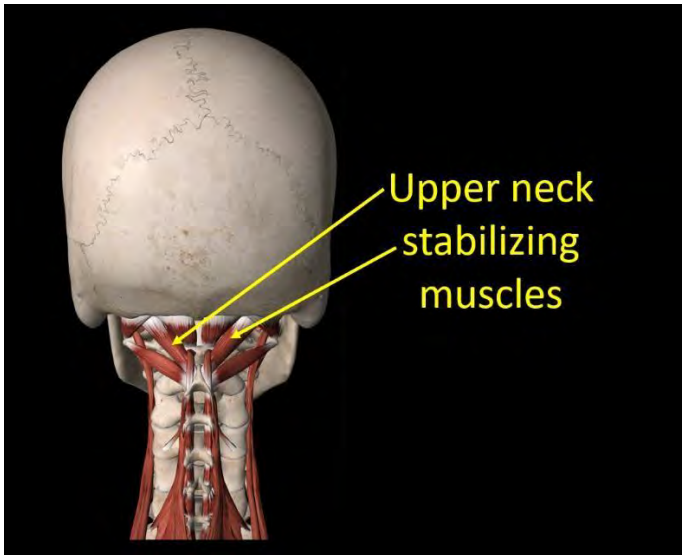
At the time of this writing, about 7 in 10 CCI patients can be helped with this approach and avoid fusion. Here are two PICL cases to conder:

- FW was a physical therapist involved in a very high-speed rear end crash where his seat back broke. He was eventually diagnosed with CCI after becoming disabled and unable to practice. His C1-C2 overhang on DMX was severe and he could do very little without severe pain. His recovery is detailed here [https://youtu.be/9Oo0v9cHdGY]. After several PICL procedures, he can now do things like ski with his daughter and hike for miles in the Colorado mountains.
- RL is a chiropractor who was in a car crash in Canada. He became completely disabled due to CCI, having to lie down 23 hours a day and only being able to be vertical for four 15-minute periods. After several PICL procedures, he was able to reverse that, meaning he can now be up for the whole day other than four 15-minmute lie down periods.

Strengthening

It’s absolutely critical that you understand that strengthening is a critical part of the PICL procedure. Why? Because half of stability is ligaments, and the other half is muscles. Hence, helping tighten and repair ligaments is only half of the treatment.

Why Do You Need to Rehab if You Have CCI? What do CCI Exercises Look Like?



Stability has two major components: ligaments and muscles. Ligaments check joints from moving too much. Many CCI patients have loose ligaments due to damage or laxity and these allow too much upper neck joint movement (typically at C1-C2 and/or C0-C1). However, what's talked about less often is that the upper neck muscles that stabilize the head on the neck also become small and weak (aka atrophied). These muscles are shown here. In order to get the most out of the CCI procedure, you have to begin strengthening these muscles that haven't worked well in months or years. The main concept is that you'll be able to do more as you become more stable.

Another strategy for higher functioning CCI patients is trying this program before the procedure. While many CCI patients can't tolerate any physical therapy exercises, some can. However, in this case, be very cautious and go very slowly. If you find you can't tolerate these exercises before the procedure, then don't despair, most patients can do them once the ligaments are tightened down by the procedure.

Level 0

A critical first step BEFORE you start any rehab program is to determine if you're ready to start at all. That's a big deal for CCI patients, as without some ligament stability this program won't be effective. While most patients would start this program several months after the first or second treatment has begun, some may be able to start early. Either way, you need to answer "No" to all of these questions if you're going to begin this program:

- Do simple head movements throw your upper neck "Out" or cause severe symptoms? Y/N
- Can just random light tasks like reaching for something or typing throw your upper neck "Out" or cause severe symptoms? Y/N
- If you use an upper cervical chiropractor, do your adjustments hold for less than just a few days? Y/N

If you have any confusion about whether it's time to start, talk to your doctor. When I refer to being "out" or a flare-up throughout this document, this means that your symptoms get worse due to the activity. For most CCI patients that would be headaches, dizziness, visual disturbances, rapid heart rate/anxiety (vagal nerve related symptoms), or neck pain.

Adjunct Therapies

If you work with an upper cervical chiropractor and this type of treatment helps, then you should continue adjustments after the PICL procedure and especially during this rehab program. While I have set this program up as something you can do at home as finding a physical therapist with CCI experience is difficult, if you have a physical therapist who has helped, then please provide him or her this book so they can see what we want post-procedure. Also, curve restoration therapies (aka chiropractic biophysics) may or may not be recommended for you, but can be continued during this program. In addition, any activities that cause you reliably to "go out" or significantly flare your symptoms should be avoided. Meaning, no pain no gain does not apply here.

In particular, depending on how much ligament stability you've gained through PICL and/or how much muscle atrophy you have as well as other things (whether irritated nerves or joints are involved), you may move this program more or less slowly than I have described. That's OK. If it takes you 2-3 times as long, that's fine. Similarly, if you can move through it quickly without flare-ups, then that's great. Again,

LISTEN TO YOUR BODY! In addition, if you can't do it at all, don't worry. You may just need more ligament tightening through another PICL procedure before this program is right for you.

One word of caution. This rehab program is NOT for patients who have already had an upper neck fusion for CCI. It should only be attempted upon the advice of your treating physician.

Level 0.25 – Getting the Right Equipment for Your Craniocervical Instability Exercises

To do these exercises, you need a LASER headlamp system. There are a few different options from low budget to medium. This is a LASER pointer that sits on top or on the front of your head while you look at where the LASER is pointing. You will use this rig to begin to get position sense and strength back in the upper neck muscles.

Medium Price

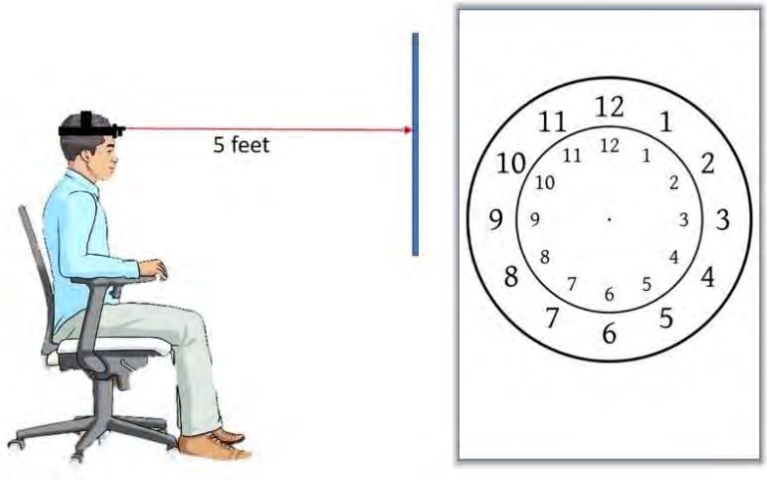
The SenMorCor system is a headlamp and wall target system that comes ready to use out of the box. It looks like this (click on the picture to the left for an Amazon link). You put that black strap device on your head with the LASER pointing forward. You then place one of the posters on the wall and use your head movements to hit various targets. The farther you are from the wall, the smaller and more fine-grained the movements. If you're closer, then the movements of your head required will be bigger. This rig and wall posters run just over a hundred USD. More links to buy this system:

- OPTP
- Physiotherapy Room

Lower Price

I found this system on Amazon which is only 20 bucks and looks like a knock off of the SenMoCor device. Some physical therapists who left reviews said it worked fine. You can also buy a wall target like this one on Amazon. You can also easily make your own as shown below, which will be required for the lower levels of this rehab program.

Level 1-Small Clock Number Movements



Now that you have your LASER pointer headpiece, find a dimly lit room. There are two options for making your exercise targets. So, let's dig in.

The cheapest way to do this is to get a 36 X 48-inch white poster board (not glossy, but matte) and a black magic marker. Draw two clock faces as shown to the left (click on the picture to see a bigger image). This is two concentric clock faces and the larger one should fill the whole width of the poster (36 inches). The smaller clock should be 24 inches wide (diameter). There should be a point in the middle of the circles. You can use bowls to draw the circles or a pin with string in the middle point and attach that to the marker while drawing a circle at that set length. Alternatively, I have designed a PDF file that you can upload to a local or online printing service.

The link to the PDF containing the CCI exercises is here [https://s5q6n6g5.rocketcdn.me/wp-content/uploads/2020/01/CCJ-Instability-Institute-CCI-Exerciseshttps://s5q6n6g5.rocketcdn.me/wp-content/uploads/2020/01/CCJ-Instability-Institute-CCI-Exercises-v2.pdfv2.pdf] . I got it printed for $25 at VistaPrint on a matte 36X48 poster. You can use any other service including your local print shop like Kinkos or office supply store (Staples and Office Depot have services). The nice part about uploading the PDF online and picking up the print is that you can save on the $9 shipping fee that Vistaprint charged.

Place the poster on the wall and get a comfortable chair. You will be measuring the distance from the wall to the headpiece and placing some pieces of tape on the floor to ensure that you know where the chair should be.

Now that you have the equipment, it's time to set up your level 1 exercise area. Place the chair 5 feet back from the wall (this distance is from your head to the wall). Get yourself aligned horizontally to the poster (you should be aligned left to right with the center of the poster and looking at it). Put the LASER device on your head and move the LASER pointer to the center point of the clock faces.

How the device sits on your head and the poster height should be such that when your LASER point hits the center of the circle, that's your neutral comfortable head position (head straight without being turned left or right or up or down). Some CCI patients have slightly off-kilter head positions that feel more comfortable, but don't worry about this right now. Hence, you may need to adjust the device on your head so that the LASER point hits the middle of the clock with your head in that comfortable

position. Make sure your shoulders are back and relaxed and that you're sitting up straight (no slouching or keeping your head forward). If this is difficult, then try a small pillow in the small of your back (lumbar) which will often help keep your shoulders back.

You should try this program as written with head movements. Again, if you can't do it without a significant consistent flare-up, then you likely need additional PICL treatment. However, you could also try an "eyes" only version of this program to start. This means you just look at these targets while keeping your LASER point in the middle of the clock face. This will still cause some very minor firing of the muscles we're interested in and may help coordinate your eyes to your cervical system before starting the part of the program where you move your head. Level 1- CCI Exercise 1

Level 1 Exercises-From the center to a number and back.

For this exercise, we'll be using the small clock face inside the bigger one. The first exercise is moving the LASER point from the center point to the 12 and back (as shown to the left). Do this very slowly. Now add in the 9, 3, and 6. So from the center to 12, back to center, to 9 and then back to center, then to the 3 and back to center, then to the 6 and back to center.

Again, these should be slow and controlled movements. In particular, as with the entire program, watch the LASER point and make it move smoothly and in straight lines from target to target. If it shakes back and forth or doesn't initially move in a straight line that's fine. But your goal is smooth movement in straight lines from point to point.

Start with what you can tolerate up to 10 reps of going to 12, 9, 3, and 6. One rep is defined as 12, 9, 3, and 6. If this is difficult for you, then you may need to stay at this level for a week or more to get to 10 reps. If it's easy, then for day 2 or 3 move onto level 2.

This is a good place to realize that you need to listen to your body VERY CAREFULLY with this program. If you have any flare-up or you feel yourself "go out" then STOP. In that case, perform fewer reps next time or move back to the easier level or exercise!

Level 1- CCI Exercise 2

For this exercise, start at the center point and then try 11, 1, 7, 5 in the way as described above (center, number, back to center). This is 10 reps. Initially just perform L1-Ex2 (Level 1, Exercise 2) alone for a few days. Then integrate L1-Ex1 + L1-Ex2 together (so 20 reps in total). You may want to stay with these first two exercises for the first week or move on more quickly if this is easy.

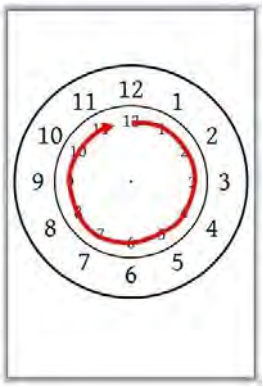
Level 1- CCI Exercise 3

For this exercise, start at the center point and then try 10, 2, 8, 4 in the way as described above (center, number, back to center). 10 reps.

Initially just perform L1-Ex3 alone but then quickly integrate L1-Ex1 + L1-Ex2 + L1-Ex 3 together (so 30 reps in total).

For week 2 at level 3, up the reps to 15 for all exercises (45 reps total). For week 3, up to 20 reps for all exercises (60 reps total). Again, if you can move through these more quickly than that. great. If you need to go more slowly than described here, that's fine as well.

Level 2 – Small Clock Circles



Level 2 – Exercise 1

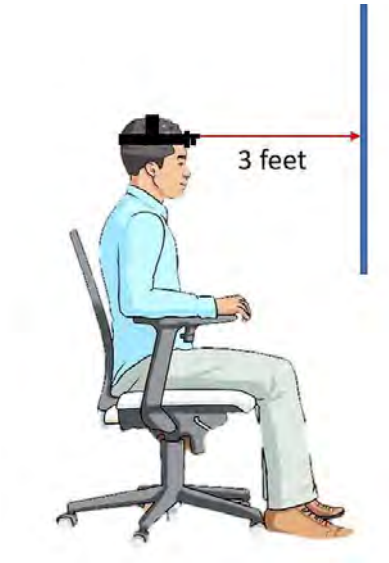
Here, start at the 12 and move the LASER pointer carefully around the clock in a circle all the way around. Hit every number starting at the 12 and going back to the 12 as shown to the left. On day 1 of this new level, do this for 10 reps, initially only this exercise (no level 1 exercises). On day 2, do 20 reps. On day 3 add back in your level 1 exercises in addition to the 20 reps of level 2, exercise 1. Keep with that program for the rest of that week. The next week add in 20 reps of the going in the opposite direction, starting at the 12 and going counterclockwise, for a total of 40 reps of level 2 exercises.

Again, how many exercises from which levels you are able to do here depends on how much you can tolerate. Higher functioning patients may find that they can do all level 1 plus level 2 exercises easily. Other patients may need more time to integrate level 1 and two exercises.

Level 3 -The Big Clock

For level 3, you should repeat level 1 and level 2 using the big outer clock face. For lower functioning patients this may take the whole time periods described for those levels. For higher functioning patients this may only represent a week.

Level 4 – Upping the Movements by Pulling the Chair Closer

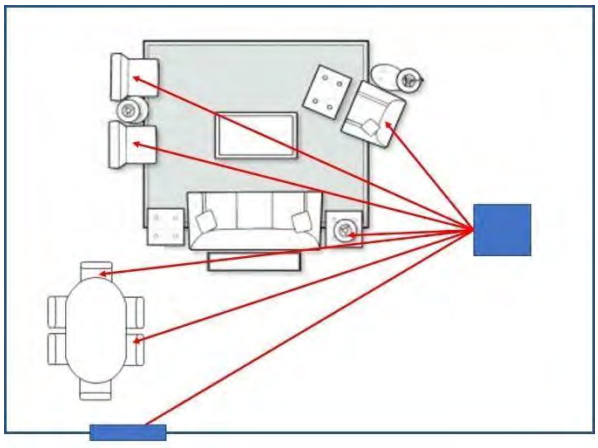


Now we're going to make things harder by moving the chair closer to the wall. The distance between your head and the wall should be 3 feet. For level 4, if this is an easy jump for you, repeat level 3 using the big clock. If this is a harder jump with more flare-ups, then repeat levels 1 and 2 before going on to level 3. This level will require more head movement than levels 1-3, so go slowly and listen to your body.

Level 5 – Closer and Closer

For this level, move the chair so that your knees are touching the wall. This should be about 18 inches to two feet depending on the length of your legs. Again, as above, if this is tough, repeat levels 1 and 2 before going to level 3. If it's easy, repeat level 3.

Level 6-90 Degree FOV Room Targets



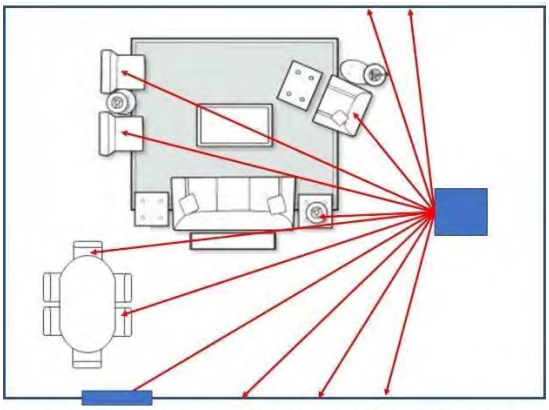
Level 6-Pick targets around the room to randomly hit with your LASER that are withing 90 degrees of your visual field.

Next, you want to begin hitting random targets around the room. In this case, make a list of the things that are within 90 degrees of your visual field (see image left). This is 45 degrees to the left and 45 degrees to the right. Make sure some are higher, and some are lower, and you have an equal number on both sides. Then write those targets down on a card and have someone randomly read them to you while you very slowly look at them and move your pointer on to them. As you get better at this level, you

can increase the pace and then more quickly go between targets. Again, start slow and slowly increase your pace.

Try this for a week starting with 20 reps and working up by the end of 1-2 weeks to 40 reps. Again, if you can do this without flare-ups, then you may want to begin timing yourself to see how long it takes to get through the list and then trying to reduce that time as the days wear on. If this is difficult, then you may want to stay at this level longer and slowly up your pace. The goal of this level is to train your muscles to be able to react to the real world where you're often forced to look at things in your environment on a moment's notice.

Level 7-180 FOV Degree Room Targets



Level 7-Pick targets around the room to randomly hit with your LASER that are withing 180 degrees of your visual field.

For this level, you increase your targets to 180 degrees (about 90 degrees to each side as shown). Do this just as described in level 6.

Level 8-Mazes

For this level, you can either make a maze on the other side of your clock poster board or buy one like the one I have shown here for $25 on Amazon. If you search google for maze patterns, there are also many you can find. In addition, the SenMorCor system I showed above has a maze poster that's included. I've also created a maze pattern that you can get printed just like the clock faces above. That PDF download link is here [link to https://s5q6n6g5.rocketcdn.me/wp-content/uploads/2020/01/CCJInstability-Institute-CCI-Exercises-Maze.pdf].

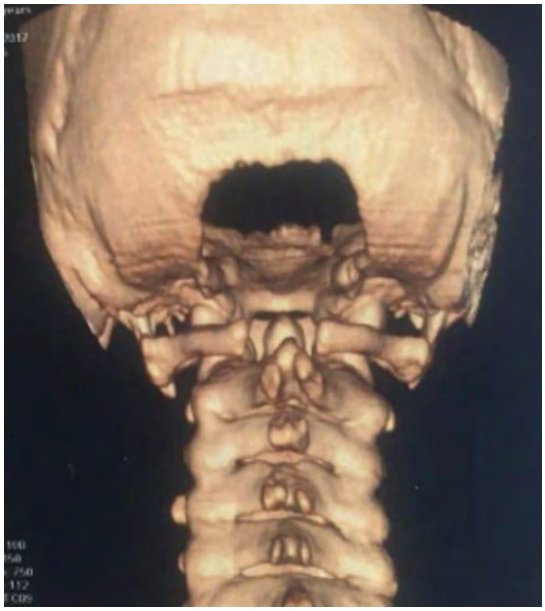
Here you begin at the beginning of the maze and move your LASER pointer though to the end. Having a few different ones to work with so you're thinking and moving your head at the same time is a good idea. Start with 20 reps at 3 and 5 feet, moving very slowly to start and then gradually increase your pace. You can also time yourself so that you can improve your times over a few weeks.

Chapter 7-Surgery

There are many different types of surgeries performed on CCI patients. The two main types are decompression and fusion. There are also secondary procedures that some CCI patients get performed that I'll also discuss like detethering and styloidectomy. Let's dig in.

Decompression Procedures for CCI

Posterior Fossa Decompression



This is a procedure where the doctor "decompresses" the back of the skull by removing bone for patients who have a low-lying cerebellum (Chiari malformation). As shown above, this is carving a hole in the back of the head.

What Is Chiari Malformation?

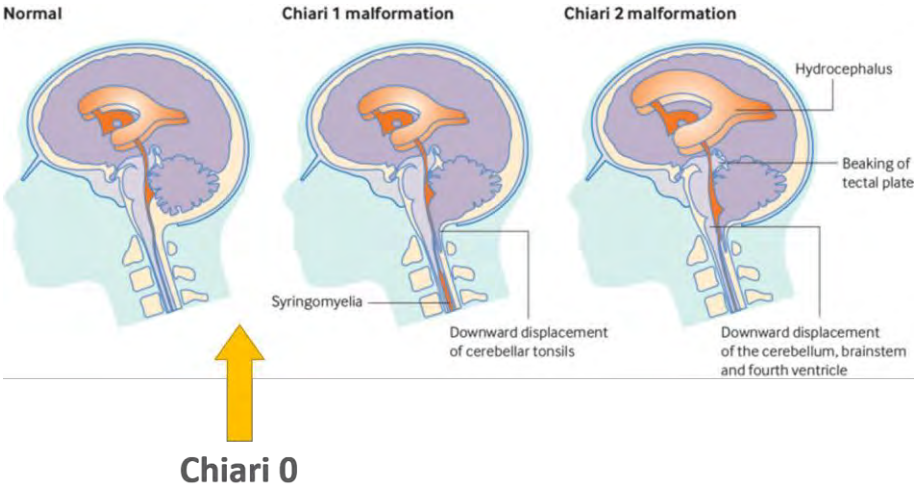
Chiari malformation occurs when a small part of the brain pushes down through the hole in the base of the skull called the foramen magnum. This can cause pressure on the back of the brain (cerebellum) and/or the brain stem. This can lead to a host of symptoms:

- Neck pain
- Balance problems, dizziness, or vomiting
- Muscle weakness or numbness
- Difficulty swallowing or speaking
- Vomiting
- Ringing or buzzing in the ears (tinnitus)

The biggest concern is that these symptoms overlap with many other problems including CCI. For example, patients with upper neck facet joint injuries get neck pain, balance issues, and other symptoms. Patients who have craniocervical instability (CCI) often have all of the above symptoms.

Hence, having an MRI suggestive of a Chiari malformation and these symptoms does NOT mean the next step is decompression surgery.

Types of Chiari Malformation



Where the rubber meets the Chiari road is which type of malformation is present. The least severe is actually not listed in the above pictures because it's more controversial, which is often called Chiari 0 or cerebellar ectopia. Few physicians would argue that if you have a portion of your brain that has herniated downwards into the spinal canal then you have a very real need for Chiari malformation treatment through surgical decompression (Chiari 1 or 2 as shown above). However, all too often these days, patients get operated on for Chiari 0 without a frank lower brain herniation, which we'll discuss in detail below.

Chiari 0 or CTE (Cerebellar Ectopia)

First, Chiari 0, or Cerebellar Tonsillar Ectopia (CTE), is when the back of the brain (cerebellum) isn't really herniating down through the bottom hole in the skull. Instead, it's "on the line" of pushing down and out of the skull or has done so just slightly. All of this is much less severe than Chiari 1, which means that surgery for Chiari 0 is VERY controversial. For example, we don't have a huge body of research that shows that this MRI finding is associated with symptoms (14).

My colleagues and I performed a research study many years ago which looked at how many cerebellar ectopia or Chiari 0 patients could be found on upright MRI imaging after car crashes (15). We found that about 1 in 5 patients with chronic pain after a car crash had this issue. The question was whether this was causing their symptoms, or an upper neck injury, as many of these patients also had evidence of CCI. Meaning that there was no way to tell whether the ligaments that held their head on or their brain suspended in the skull had been injured and the Chiari 0 was being caused by that ligament injury.

Diagnosing Chiari 0

This is a bit of a sticky wicket. Meaning that we have no sure-fire way to determine if what we see on your MRI as Chiari 0 is in fact causing your symptoms. This is a big problem.

For example, if we suspected you had pain in the right C1-C2 facet joint I could inject numbing medicine using radiographic contrast confirmation and x-ray guidance to make sure that I was in the joint. If your

pain and symptoms go away, we have our answer, which is that this specific joint is causing your problems. However, I can't do that with Chiari as there is no safe way to numb out your Chiari malformation.

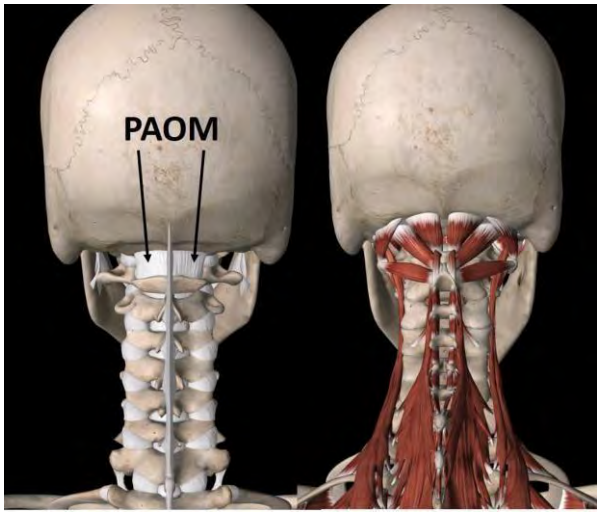
The only way that is used to try to diagnose Chiari is to use invasive traction. This is where the doctor places screws into the skull and pulls upward, trying to make space for the lower brainstem and to check if your symptoms go away. The problem? This also takes weight off of the upper neck joints, discs, and spinal nerves. Hence, the pain could be coming from any one of these areas. Hence, the only real way to see if Chiari malformation is causing your symptoms is to cut a hole in the back of your head (decompression) and see if this makes the symptoms better. Surgery for Chiari 0

Chiari malformation treatment often involves surgery. The goal is to remove bone and "decompress" the area of the brain with pressure. That means opening the skin, retracting the muscles, and then removing the bone shown here in yellow. This is often the back of the skull and the back of the c1 and/or C2 vertebrae (if needed). The skull bone removal procedure is called a posterior fossa decompression.

Complications from these procedures are substantial (16). They include more than 1 in 10 of the patients having a dural leak. This means that the doctor inadvertently cuts through the covering of the brain and spinal cord (the dura) which is the sac containing cerebrospinal fluid in which the brain floats. The dura is then sewn back up and leaks, often requiring another surgery. Other common complications include 9% who get an infection of the brain (meningitis), infected wounds at 7%, and damaged nerves in 5%. **About 3% of the patients die from the surgery.**

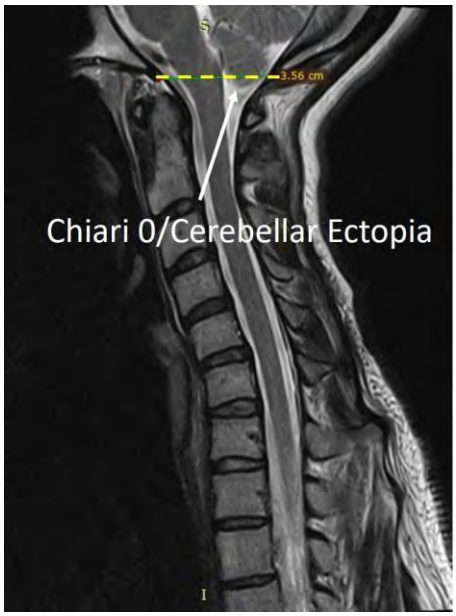
What Is the Collateral Damage?

If you survive the surgery, a common issue is the collateral damage. To understand that we need to review the critical anatomy in this area. In the image shown here, note that the PAOM is a ligament in the back of the neck that helps to keep the skull stable on the neck as you look down. Then note the myriad of muscles in the upper neck at the base of the skull. They're responsible for maintaining the stability on your neck. Both this ligament and many of these muscles are destroyed when a posterior fossa decompression surgery is performed. Why is this a big deal? Permanent Bobblehead



This ligament and these muscles at the back of the upper neck and skull are critical for stabilizing your head on your neck. Think of the head as a bowling ball at the end of a stick (your neck). Without an important piece of duct tape (the PAOM ligament) or the muscles that actively balance the bowling ball on the stick, you can become a permanent bobblehead. Hence, outside of huge fusion surgery to bolt all of this together, there is no way to fix the collateral damage done by this aggressive surgical Chiari malformation treatment.

My Patient



This is my patient's MRI. It shows a Chiari 0 or cerebellar ectopia, meaning that she doesn't have a Chiari 1 with a good chunk of her lower brain having herniated through the bottom of the skull and into the spinal canal. Instead, the surgeon believed that pressure on the lower part of the brain (the cerebellar tonsils) was causing her symptoms. She therefore underwent a posterior fossa decompression at great personal risk. This didn't help any of her symptoms which primarily include balance problems, neck pain, and brain fog. She was then later diagnosed with craniocervical instability (CCI) by a neurosurgeon in

Spain who wants to now fuse C0-C2 (the skull to her upper neck bones) and her lower neck to boot. She wisely said no and began to explore other options.

Fusion

Surgical fusion is usually the absolute last resort procedure for any patient with a spine problem and this is doubly true for CCI patients. There are two common types of upper cervical fusion:

- C1-C2 Fusion
- Occipital-cervical Fusion

Let's focus first on the most common type of fusion surgery applied these days in CCI patients, a C1-C2 fusion.

C1-C2 fusion means using screws, rods, or bone to make sure that the C1-C2 joint doesn't move. While the most common reason this rare procedure was performed used to be because of an upper neck fracture or severe ligament rupture, these days more of these procedures are performed in patients with ligament laxity leading to upper cervical instability. This is where this review of C1-C2 fusion will focus.

C1-C2 Screw Fixation or Magerl Technique

C1-C2 Screw Fixation-A single screw is placed across the C1-C2 joint.

The two most common types of C1-C2 fusions that I see in patients I consult on are posterior screw fixation (Magerl Technique or C1-C1 trans-articular screws). In the C1-C2 screw fixation, a single screw is placed across the joint as shown here. The positives for this technique are that it disrupts less of the stabilizing muscles at the back of the upper neck, which are critical for stability. Since the goal of the procedure is getting the normally mobile C1-C2 joint to grow together and become one solid mass of bone, when this doesn't happen, it's called a non-union.

Non-unions are a big problem that I see in patients who have had this procedure. They are then left with a screw through their joint, which destroys it. Without a fusion, that screw moves as they move their head, which further damages the joint.

Based on the published literature, an even bigger issue seems to a risk of vertebral artery injury (8). This is the artery that supplies blood to the back of the brain. An injury here requires emergency vascular/neurosurgery and can be life threatening.

Goel and Harms C1-C2 Fusion Technique

Goel and Harms Technique-Two bone screws and a rod are used, with or without the addition of bone graft.

The Goel and Harms technique shown here is more extensive than the C1-C2 screw fixation. Here two screws are placed into the bone and a rod system is used to hold the C1-C2 joint. Bone chips from the patient are often placed around the rods to help provide stability as the bone grows. The big downside with this approach is the extensive muscle damage that's required to get all of this hardware in place. Other risks include again, damaging the vertebral artery. In addition, many surgeons will also sacrifice the C2 nerve, which can lead to chronic headache pain.

Occipital-cervical Fusion

In this surgery, as shown here, the base of the skull is fused to the upper neck with rods and screws. This procedure has very high complication rates of about 50% (8). Also, fusing the upper neck to the skull places tremendous pressure on the remaining neck bones, which is a set-up for adjacent segment disease. That means that the levels below the fusion wear out due to the extra pressure, often requiring more surgery.

Upper Cervical Fusion Risks

Now that these procedures are being performed for non-life-threatening injuries like ligament laxity, the risk-benefit ratio is different. For example, when a patient with an upper neck fracture may have died or become a high quadriplegic if the fracture wasn't stabilized, the risks allowed for the surgery can be quite severe and the risk versus benefit equation still makes sense. However, now that these invasive

procedures are being performed for patients with loose upper neck ligaments due to damage or congenital problems like EDS, the risk-benefit bargain can be a bit off. Let me show you what I mean.

Nonunion

One of the big risks of any fusion surgery is that the joint being fused never actually grows together. For these procedures, accurate information that applies to adults is hard to find, as many of these surgeries are performed on children with congenital abnormalities. However, at least one author states that nonunion rates can be high with these techniques (9). However, there's nothing like a good case report to make complications personal. Katie is a twenty-something I treated several years ago. She had a C1C2 screw fixation performed for CCI after a DMX showed too much movement at C1-C2 and nothing else was helping. Regrettably, the joint never fused, leaving her with new strange movements of the C1-C2 joint as it pivoted around the screw going through it. The joint was also damaged by the screw and still moving, making her headaches worse and not better (pain from the C1-C2 joint refers to the head). She was also not a good candidate for our PICL injection procedure due to the surgery. Through injections of PRP into the C1-C2 joint, we were able to get the joint to fuse, but she also had damage to the occipital nerves and extra force across the C0-C1 joint leading to new pain there. These areas were treated with platelets and stem cells with some improvement.

Misguided Screws

A big issue we're seeing in the clinic is the fact that these screws placed into bone are hard to guide. Hence, they can inadvertently reach places that can damage structures. For example, the screw can hit the vertebral artery and damage it, end up hitting and damaging nerves, and even destroy the C0-C1 joint. Since these screws are large, what they hit is usually obliterated. The vertebral artery runs through the neck bones and this upper neck area. It supplies blood to the back of the brain.

The good news is that most people have one on each side, so losing one vertebral artery can often be compensated for by the other. Realize though, that destroying one of these arteries with a big screw is still a very bad thing. For example, in an older patient, this could lead to a stroke (blood clot or other debris floating into the blood supply of the brain). This happens 4-10% of the time with upper cervical fusion (8,10). At the very least, emergency vascular surgery may be required.

Damage to the upper spinal nerves can also happen (11). The most common nerve injury is to the C2 spinal nerve (3). This supplies the back of the head, so damaging it can lead to chronic headaches. As discussed above, the C2 spinal nerve is also sometimes sacrificed in the surgery itself. That means it's taken out by the surgeon because it's in the way of the desired screw placement. This can also lead to chronic head pain about 1/3 of the time.

The destruction of the C0-C1 joint is also possible and frankly, one of the most common complications I have seen. Here the screw is placed too high and travels not only through the C1-C2 joint, but also through the C0-C1 joint, which destroys the joint. This means that the screw causes arthritis in the joint, which is bone and cartilage damage. Hence, the C0-C1 joint becomes a new source of pain. This joint also causes headaches at the back of the head.

Complication Case Reports

The first patient is Rosalyn, who was a student at an elite university. She was diagnosed with CCI due to a car crash and had moderate instability on her DMX and evidence on MRI of alar/transverse ligament

injury. She underwent C1-C2 screw fixation where a screw was placed through the C1-C2 joint to fuse it and to reduce her instability. Regrettably, on one side that screw was placed too deep and she woke up from surgery with new pain right at the base of the skull on that side. This worsened over the ensuing few days and she also reported a “scraping sound” at that spot when she went to look up or down. That was the screw scarping off some of the cartilage in the C0-C1 joint. She then underwent subsequent surgeries to remove the screw, but by then she had two problems. The first was severe arthritis in this joint and disabling new headaches from that damage. The second was that the two surgeries destroyed most of the critical stabilizing muscles in this area, which only increased her instability. We tried to perform a PICL injection procedure and injected stem cells into the C0-C1 joint to aide repair, but this only helped minimally as the damage from the screw was extensive. The good news is that as far as I know, she was able to get enough relief to finish college, but the bad news is that she continues to have this disabling pain to this day.

C1-C2 Fusion

The second case is also of a young woman, but her issue was a loose C1-C2 level due to EDS. She had a modified Goel and Harms technique C1-C2 fusion. On the right, the screw was placed too high into the C1 bone and ended up inside the C0-C1 joint. She reported immediate pain at that site after surgery. However, it seems that she wasn’t taken seriously and it took 3 months to get imaging showing the problem. The screw was eventually backed out, but then the mass of bone placed in this area to promote fusion fractured, leaving her without any bone stabilization. On a positive note, all of the new scar formation from the surgery provided some stability, but the recommendation is still to fuse her from C2 to the skull, which she didn’t want.

Adjacent Segment Disease

Adjacent segment disease (ASD) is the bane of every fusion. ASD happens because all spinal levels are built to move just a little bit. When that movement is stopped due to fusion, the levels above and below take too much force and can develop degenerative arthritis and breakdown. Here the C1-C2 joint is responsible for half of all of the rotation of the head on the neck, so fusing it dramatically increases force both on the C0-C1 and C2-C3 joints above and below. Meaning that over time, you can expect these levels to break down in most patients. Far too many of these patients, in my experience, will eventually require additional surgical fusions above and below.

The Other Side of the Argument

There are patients who really need these surgeries. They are few and far between, but they are out there. In fact, I can remember one young woman from more than a decade ago who had life-changing results. She was completely 100% disabled and had a hard time doing anything but lying down. She had an upper cervical fusion and was able to begin functioning and even have a child.

Having said that, I’m seeing much higher complications and side effects than have been reported by surgeons in the literature. Hence, trying less invasive procedures first just makes common sense. Again, remember the ladder of invasiveness and use it as your guide!

Who might consider fusion?

- There is an active fracture in one of the upper neck bones

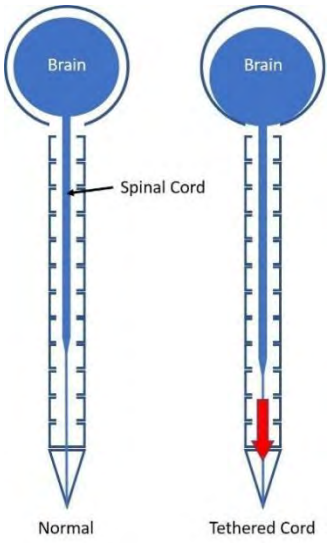
- Conservative care and PICL have failed to help
- The patient is not a candidate for PICL, but has diagnosed CCI
- The level of Chiari malformation or a physical obstruction like a pannus leading to cervical medullary syndrome is severe and leading to direct compression of the spinal cord

The upshot? Upper cervical fusion is not a routine spinal surgery. It's much higher risk than the average procedure due to the vertebral artery as well as the other problems I've shown. In addition, I've seen far too many misplaced screws. So, while there are a handful of patients that really need this surgery, please do your homework!

Detethering

This is a surgery to cut the connective tissue that anchors the end of the spinal cord to the spine. It's performed for patients who have an occult tethered cord. Let's dig in.

What Is a Tethered Spinal Cord?



At its most basic, your spinal cord projects down from your brain and contains the major wiring that tells your muscles to move and allows you to feel things. It's connected to the brain and travels through the spinal bones (vertebrae) and usually ends at the lowest part of the upper back (T11-L1). There are nerves below that are called the cauda equina (horse's tail). There's also a piece of connective tissue that anchors it from below called the filum terminale (filum). In addition, there are a ton of small ligaments throughout the cord that help to anchor it all the way down.

When something from below causes the filum to get stuck, like a tumor or scarring from prior low back surgery, the filum can yank on the cord and pull it down as shown here. When that happens, it's called a "tethered cord" (17).

Most of the pressure is on the lower part of the spinal cord with the connective tissue of the cord diffusing the pressure as you go up. That's why most of the symptoms are in the lower back and legs (lower spinal cord and nerves). However, another aspect of that diagram is that some believe that a tethered cord can also pull on the brain and cause it to hang low in the skull base, intersecting with a disease called "Chiari Malformation". More on that below.

The most frequent tethered cord patient is usually a kid with Spina Bifida. It can be congenital (the person is born with it) or acquired later in life. If it's the latter, there is usually something pulling on the filum terminale or it's stuck on a physical obstruction or scarred down. That obstruction can be a fatty tumor, a spinal cord tumor, or a bone spur. In addition, local scarring of the nerve roots after back surgery (i.e. failed back syndrome) is also a known cause. In rare instances, this problem can also be seen in scoliosis.

Symptoms

Symptoms include back pain that radiates to the legs, hips, and genital or rectal areas. There can also be bowel and/or bladder issues. The legs can feel numb or tingly and, in some patients, get weak or begin to lose muscle.

Diagnosis

This is where you need to break an adult with a tethered cord into two camps: an obvious tethered cord and occult tethered cord. For an obvious tethered cord due to masses in the spinal canal or prior back surgery, the diagnostic criteria is that the end of the spinal cord which is normally located at T12 or L1 is low, below L2 with a thickened filum. However, for occult tethered cord, there is no mass or prior back surgery, the diagnosis can also be made on clinical symptoms, abnormal urodynamics (testing), and a lack of other things helping the condition.

Urodynamics

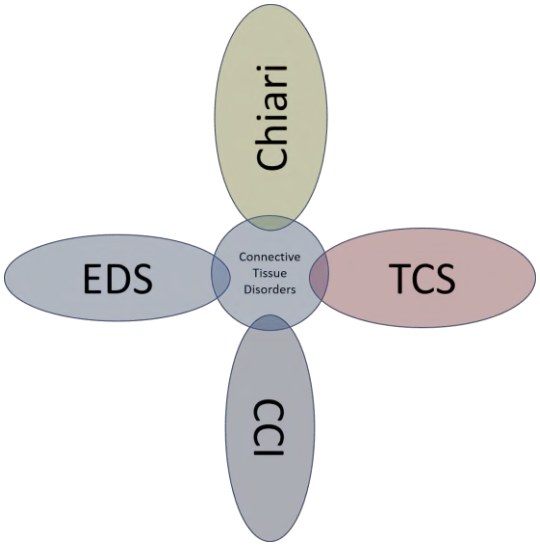
One of the items that occult tethered cord and detethering advocates point to as an indication for the surgery is urodynamics. This is testing of the bladder performed by a urologist that can identify a "neurogenic bladder". The concern I have is that we have seen many patients through the years with low back pain and sacral nerve irritation with a neurogenic bladder on urodynamic studies. These are commonly patients with a central L5-S1 disc bulge that irritates the descending sacral nerves or chronic SI joint syndrome that irritates the sacral nerves. Their symptoms and urodynamic studies normalize after a simple caudal epidural to reduce this nerve inflammation. So, this usual cause of a neurogenic bladder would need to be ruled out in surgical detethering candidates before a procedure was performed.

Surgery for Tethered Cord

The most common surgery for tethered cord or occult tethered cord involves cutting the anchoring tissue on the bottom called the filum terminale. This is called "detethering". Complications include infection, bleeding, and damage to the spinal cord, which may result in paralysis or loss of bowel or bladder function.

Tethered Cord in Adults

So, if a tethered cord is a diagnosis normally found in kids with spina bifida and adults who have rare spinal tumors or prior back surgery gone bad, how is it that some patients who fit none of those descriptions are now getting surgery to treat this issue?



The diagram above shows how various problems may intersect with tethered cord.

The connection between occult tethered cord surgeries and adult patients starts with Chiari malformation (18). This is a problem where the brain is hanging low in the skull and the bottom part (the cerebellum) can get pushed into the spinal canal. The idea is that the cord and thus the brain is being pulled downward. This is still pretty controversial, as the biomechanics of the spinal cord don't necessarily support the possibility that this is possible. Meaning that any downward force from a tight filum would be absorbed by the lower spinal cord as the structure is elastic like a rubber band.

Despite this, there are patients (and a handful of neurosurgeons) who believe in the diagram seen here-. That is that a tethered cord is pulling the brain down, thus leading to Chiari malformation thus these patients require detethering. That interfaces with EDS (Ehlers Danlos Syndrome) patients, who have super stretchy ligaments, because it's believed that the connective tissue that normally anchors the brain and holds it up is too loose, thus allowing it to hang low in the skull (19). In addition, these same stretchy ligaments also cause CCI and the extra movement causes irritation of the brain and spinal cord. Hence, the rationale is that they too may need detethering to reduce the downward pull on the brain.

So how do we get from CCI (craniocervical instability) to detethering? Another way we can get there is if a surgical fusion tries to correct vertical instability. The idea here is that the CCI causes "cranial settling" which is that the skull is pushed into the upper neck. The surgeon uses traction during fusion surgery and thus fuses the patient in a position where the skull is higher than normal. This then stretches the spinal cord too much, leading to the need for detethering. This is a VERY controversial topic, so detethering after surgical fusion is not a common procedure.

Research?

I searched the US National Library of medicine for any research on the following topics:

- Adult Tethered Cord Syndrome-Not much on PubMed using that search, but more on Google Scholar-A case series that again doesn't apply to this discussion as the detethering was performed for traditional causes of tethering like tumors (20). Or an overview paper that again focuses on kids or adults with spinal masses or prior back surgery (21). Or a theoretical paper on this actual topic (adults with symptoms without known masses causing tethering) but provides

no higher-level clinical data. Another paper describes 24 patients treated over 11 years who didn't have masses and had a primary presentation of back pain (22). In reviewing many of these listed papers, most again focus on patients with traditional causes of a tethered cord and not on adults with no known risk factors who suddenly develop symptoms.

- Ehlers Danlos Syndrome and detethering – NONE
- Chiari Malformation and detethering – Some, no high-level research, most in kids with spina bifida or other common causes of tethered cord like tumors.
- Craniocervical instability detethering – 2 hits-one case report on a rare congenital defect and a surgery planning study for C1 screw placement. Meaning zip on an adult with CCI getting a detethering procedure.

What Could Go Wrong?

This is the mantra that every patient considering an invasive surgery like cutting the connection of the spinal cord needs to consider. Almost all of the research on detethering and possible complications is from studies that reviewed the results in kids with spina bifida. Unfortunately, most of these kids don't walk and have severe functional and developmental delays. Meaning that detecting complications in this group would be very tough. In an adult who walks and talks and otherwise isn't wheelchair-bound, the significance of complications that could include never walking again is a much bigger deal. However, we have no real reports of the complications of detethering in this new group of patients getting the surgery.

Should You Get Chiari or Detethering Surgery?

These are NOT first-line treatments. Cutting the connection for the spinal cord is a big deal. One of the big problems that I see is that the filum terminale is there for a reason. It anchors the spinal cord and nerve roots. Cutting it will permanently impact the biomechanics of how your spinal cord and spinal nerve roots move. Once that's done, there is no going back and reconnecting it.

Styloidectomy-Eagle Syndrome Surgery

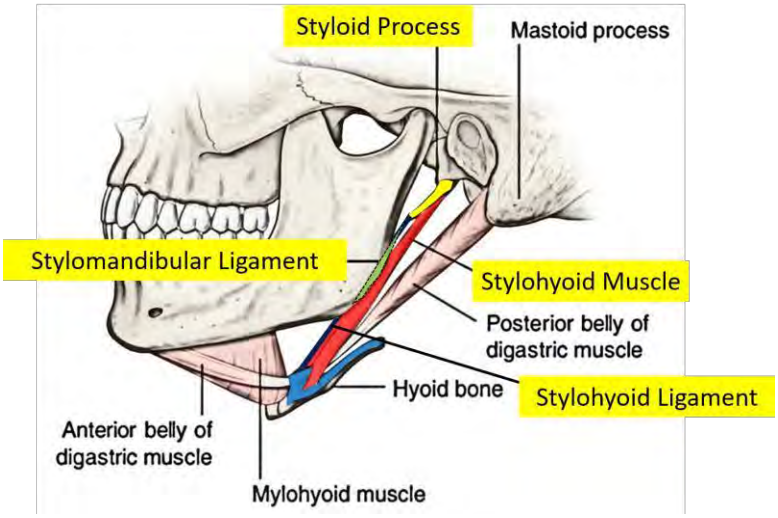
We've been noticing more CCI patients who get surgery for Eagle syndrome. What is that and why? Let's dive in.

What Is Eagle Syndrome?

Eagle syndrome was first described in 1949. It can happen when a bone at the base of the skull (the styloid process) grows too long and impinges on important nerves and/or blood vessels. The symptoms include throat pain (which can range from dull and nagging to severe) or trouble swallowing. There can also be neck and/or facial pain. This can also be felt in the jaw or ear. Other symptoms can also include tinnitus or a ringing sound in the ear or increased symptoms with head movements, such as turning the head or chewing.

This problem impacts women more than men. This is a RARE condition with an incidence of 4-8 per 10,000 people.

Anatomy



The styloid process is the key piece of anatomy to understand Eagle syndrome. This is a tooth-like projection from the side at the bottom of the skull just in front of and down from the ear (shown in yellow here). It connects to the stylohyoid muscle (red) which connects to the hyoid bone (blue). The styloid process also has muscles that go from there to the tongue (styloglossus) and back of the throat (stylopharyngeus).

There is also a ligament connecting the styloid process to the hyoid bone called the stylohyoid ligament and one that goes to the jawbone (stylomandibular ligament). Confused yet? You should be, this is a complex area. But here's the summary so far – this styloid process is connected to lots of critical stuff!

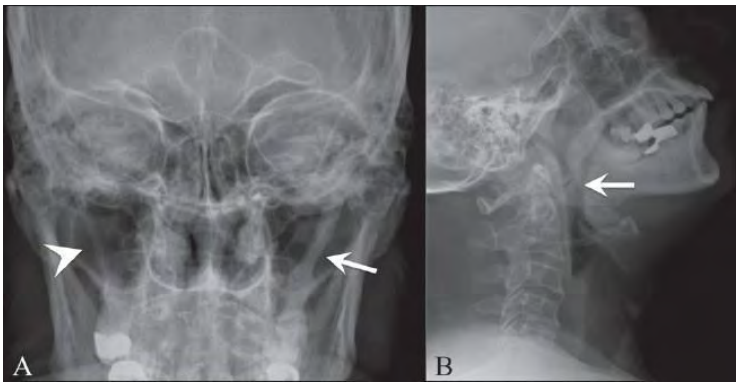
What else is in this vicinity? There are vessels and nerves in this neighborhood as well. The internal jugular vein, internal carotid artery, and glossopharyngeal nerve (CN IX), vagus nerve (CN X), and accessory nerve (CN XI) run inside of the styloid process. The occipital artery, hypoglossal nerve (CN XII), (CN VII) runs to its outside. The trigeminal nerve (CN V) is also in this general vicinity.

Why the Symptoms?

The idea here is that the elongated bone aggravates nerves in the area. Here's what advocates for this diagnosis believe happens:

- Tongue spasm and throat pain-irritation of the glossopharyngeal nerve which supplies the tongue and the back of the throat and the hypoglossal nerve which moves the tongue.
- Neck pain-irritation of the accessory nerve which supplies the trapezius muscle
- Facial Pain-irritation of a part of the facial nerve or the trigeminal nerve

How is This Diagnosis Made?



The first thing that's usually noted is a long styloid process on x-ray. A normal one is about an inch long (25mm). A styloid process that's longer than 30mm is considered abnormal. However, this is where you as a patient need to be VERY CAREFUL! Why? 4-7% of everyone walking around out there has a long styloid process, only 4% of patients with elongation of the styloid process show symptoms!

Isn't there a highly accurate test that doctors perform to tell if I'm one of those 4% of patients with a long styloid bone that is causing my symptoms? NO. Most patients are operated on based on the x-ray (or CT scan) and the symptoms.

There is an ultrasound or x-ray guided diagnostic numbing injection that can be performed, but nobody is sure if that's a 100% accurate way to choose patients who will respond to surgery. However, you should consider getting this done by THE RARE QUALIFIED EXPERT who understands how to perform this injection. If it's a positive block, your pain and symptoms should go away for several hours.

Looking at the Internal Jugular Vien

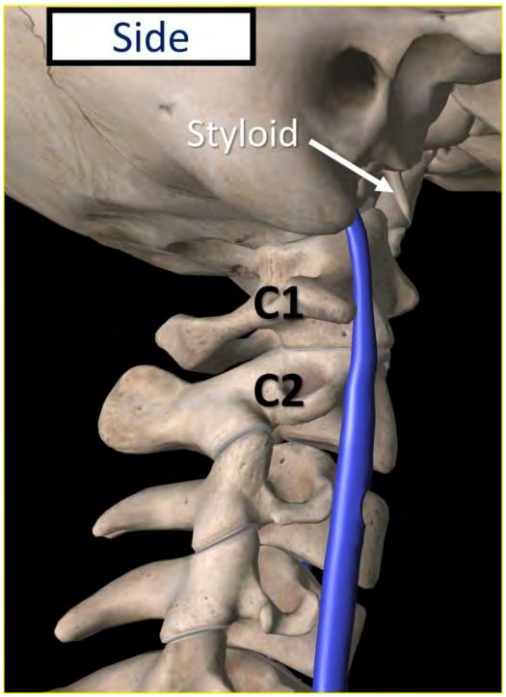
Rather than focusing on the styloid, another way to look at Eagle's syndrome is to image the internal jugular vein. This can be done using a CT venogram or ultrasound.

A CT venogram (or sometimes a CT arteriogram) is a diagnostic test where the radiographic contrast dye is injected into a vein (or artery) and then a CT scan is obtained. This can show the internal jugular vein and if it's compressed. An ultrasound of the front of the neck can also be performed, but this technique would generally be considered less accurate due to the technical demands of getting the right area imaged.

Why Could the Status of the Internal Jugular Vein Be Important?

In this context, intracranial hypertension is the backup of blood or cerebral spinal fluid in the brain. This can happen when the internal jugular vein is compressed as that could have some cross-over with neck problems.

The internal jugular vein (IJV) is in the neck and lives in the carotid sheath which also contains the internal carotid artery and the vagus nerve. The IJV takes blood from the brain and in our context is important because it passes in front of the upper cervical spine as shown above. In fact, it passes right in front of the transverse process of C1. That's the little side projection coming out of C1 and located just behind the vein as shown.



While there are many opinions about the symptoms that patients get when the internal jugular vein is compressed, let's see what the research shows us. There is a study that was purposed to measure symptoms in known cases of internal jugular vein compression (29). I took the main table of results and turned that into the word cloud you see above. Here, the bigger the word, the more often patients with IJV compression complained of that symptom. As you can see, the top three symptoms were:

- Insomnia
- Tinnitus (ringing in the ears)
- Head noises (often a "whooshing" sound)

Then came hearing problems. Interestingly, the two symptoms that many of my CCI patients believe are due to IJV compression (headache and neck pain) were the least commonly reported.

Is IJV Compression Causing Symptoms?

This is a hard one for patients to understand, but critical in helping to keep them safe. A valid test is one that's unlikely to be found in patients without symptoms and is only usually found in patients with symptoms (or the disease in question). Since the question here is whether the IJV compression is causing symptoms, then we should only very rarely find IJV compression in patients without symptoms. **However, that's NOT the case.**

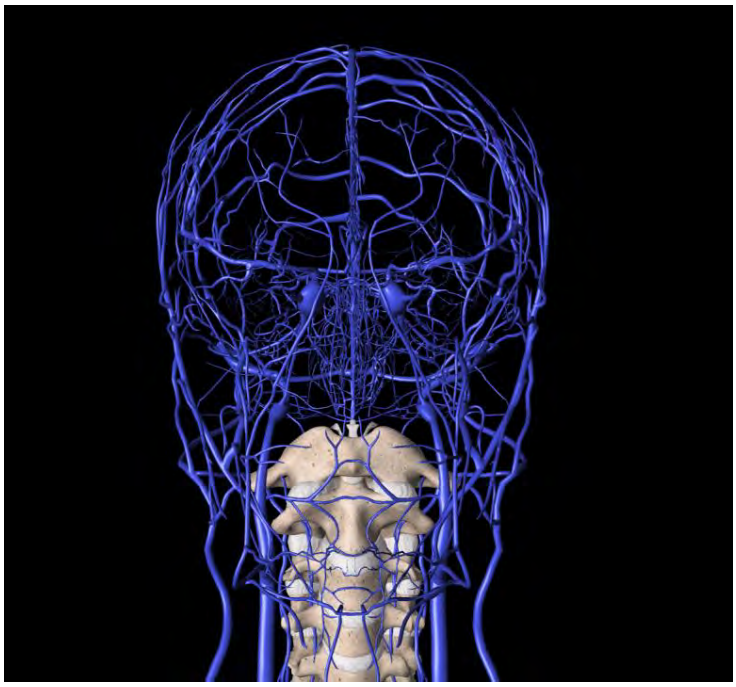
Take for example this recent study (2). The researchers wanted to determine how often IJV compression happened in patients without any symptoms. They did that by going back to look at CTA studies from patients who were usually being worked up for carotid artery stenosis. Meaning in these cases, finding IJV compression would be "incidental". In other words, it would be just a random finding not linked to any IJV compression symptoms.

How often was random IJV compression without any symptoms found? Way too often for my comfort:

- **Moderate stenosis** was seen in **33.3%/25.9%** (right/left) internal jugular veins.
- **Severe stenosis** was seen in **24.1%/18.5%** (right/left) internal jugular veins.
- Severe **bilateral** extrinsic compression was seen in **9.3%**
- The most common causes of extrinsic compression included the **styloid process** and the posterior belly of the digastric muscle.

This is a SERIOUS PROBLEM for the idea that we can use imaging that shows IJV compression to diagnose the cause of someone's symptoms. You may be asking yourself, how is it possible that these patients had compression of the IJV without symptoms? One word provides that answer, "collateralization".

What is Collateralization?



Above my diagram shows just SOME of the venous routes out of the brain. As you can see, if one route is blocked, your body can enlarge another one. In addition, your body can also grow new veins around the compressed area.

If My IJV Compression and Intracranial Hypertension Aren't Causing My Headache, then What's the Cause?

There's a long list of neck structures that, if damaged or irritated, can cause headaches. These include:

- C0-C1 facet joint
- C1-C2 facet joint
- C2-C3 facet joint
- Greater occipital nerve
- Lesser occipital nerve
- Third occipital nerve at C2-C3

- Third occipital nerve at posterior C2
- Superficial cervical plexus at the SCM
- Supraorbital and supratrochlear nerves
- Auriculo-temporal nerve
- C2 dorsal root ganglion
- C2-C3 Disc
- Sphenopalatine Ganglion
- Many muscle trigger points

Why Is My Styloid Bone Long?

Nobody is 100% sure why this happens. However, looking at the diagrams above, note that the styloid process is attached to the jaw through ligaments and muscles. Hence, chronic TMJ or neck issues causing too much force on the TMJ can pull on the bone causing it to get beat up and lengthen. This wear and tear can also cause the normal pliable ligament to turn to bone (ossify).

Do Other Things Cause These Symptoms?

YES. Many other conditions cause these same symptoms. Meaning that this 4% of patients with a long styloid bone and symptoms may actually have something else causing their pain. While I know that this may be a disconcerting message to hear when you believe you have finally found the cause of your symptoms, it's a very critical one to absorb.

One of the biggest overlaps we see is Craniocervical Instability (CCI). All of these same cranial nerves can get irritated not by a long styloid bone, but by loose ligaments that hold the head causing too much movement of the skull on the spine. This can cause irritation of these nerves where they exit the skull. Hence operating on the neck and taking out a piece of this bone will do nothing to relieve these symptoms.

We have also been tracking a number of patients who got this diagnosis and ended up with surgery. Why? As a clinic, we have seen a rash of patients getting their styloid bones removed who are no better or worse after the procedure.

What Else Could Be Causing My Symptoms?

Many things cause neck pain that are MUCH more common than Eagle syndrome, including damaged neck joints, pinched neck nerves, loose ligaments, irritated muscles, etc.... Other things cause facial pain such as TMJ syndrome and trigeminal neuralgia. Other things cause throat pain including local lesions in the throat.

Does Styloidectomy Surgery Work?

The real answer is we really don't know for sure, as there are NO gold-standard randomized controlled trials comparing surgery to no surgery. However, there are some lower-level studies that suggest that surgery may help. We do know that the surgery, because of all of the nerves and vessels near this area, has potential complications. These include:

- A localized infection requiring IV antibiotics and/or additional surgery

- Trigeminal neuralgia (damage to this nerve that causes more face pain)
- Vascular injury leading to severe bleeding and requiring microsurgical repair of the vasculature.
- Facial paralysis due to injury of the facial nerve

Are There Other Ways to Get Me Out of Pain?

USUALLY. Meaning in our experience, most patients who have been told they need surgery for Eagle Syndrome actually have other causes of their symptoms. Many different neck treatments can be applied to help these symptoms if the actual cause is a neck problem in the spine and not the long styloid process. Again, realize that 96% of patients with a long styloid process do not have Eagle syndrome.

A Surgeon Told Me that I NEEDED to Have this Removed

This messaging of severe problems like a stroke or other issue that could happen if the styloid bone is not cut out are surgical sales techniques. While for some patients with severe disease there may be additional risks, in our clinical experience, for the vast majority of patients who are told they need surgery, the risk of removing the bone is greater than the risk of stroke or puncture of vessels due to the elongated bone.

Can a Styloidectomy Surgery Impact the PICL Procedure?

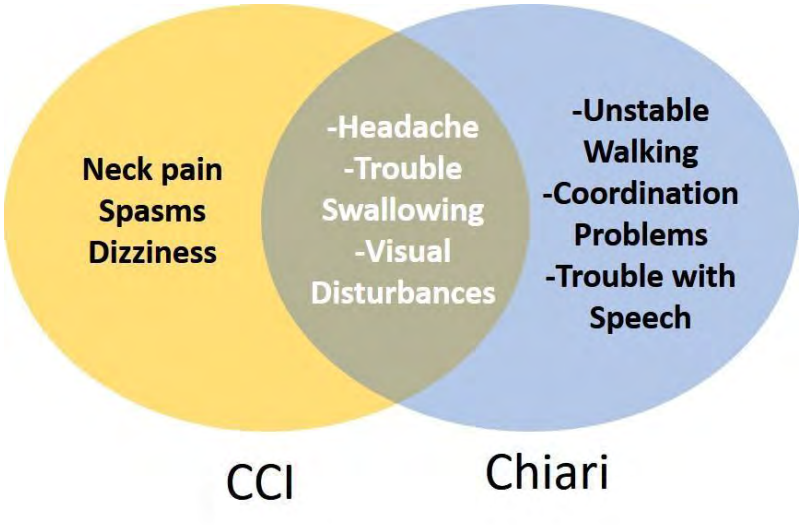
Yes, a styloidectomy surgery destabilizes the anterior neck. Why? Because ligaments and muscles connect the skull into the hyoid bone which acts as the gatekeeper of anterior neck stability. Removing this connection can cause the front of the neck to lose stability. Since CCI patients need as much stability as possible, adding more instability isn't a good thing. We have also seen a significant number of CCI patients get worse after styloidectomy surgery.

Chapter 8-Other Issues Surrounding CCI

There are a number of medical conditions that intersect CCI. What I mean is that either CCI patients tend to get these diagnoses as well or that people prone to CCI can have these issues. I've already mentioned a few under the surgical section and we'll also get into some others here:

- EDS
- Chiari malformation
- Tethered cord
- Eagle syndrome
- Dural leaks
- Chronic fatigue syndrome (CFS or ME-CFS)
- Mast Cell Activation Syndrome (MCAS)
- Postural Orthostatic Tachycardia Syndrome)
- Retroflexed dens
- COVID
- Lyme Disease
- Mold/CIRS

The hard part here is that many of the symptoms of these medical conditions overlap with CCI symptoms. For example, in the diagram below I list some of the common symptoms of CCI and Chiari:



As you can see, there are shared symptoms between these two diagnoses. A careful review of the symptoms would tend to push someone toward one or the other. In addition, the diagnosis of CCI could be bolstered by imaging. However, one of the tests to diagnose Chiari malformation is invasive traction. This is where screws are placed into the skull and the doctor pulls traction to clear space for the Chiari malformation and asks the patient if certain symptoms have improved. However, realize that if the headache pain was caused by an upper neck facet joint injury, which is common in CCI patients, since we're taking weight off that painful joint, the headache would also be relived having nothing to do with the Chiari malformation. Hence, it's important to be very careful about figuring out what's wrong by looking at only symptoms.

Chronic Fatigue Syndrome

ME-CFS or myalgic encephalomyelitis/chronic fatigue syndrome is a condition where the patient has extreme fatigue. Rest usually doesn't help, and physical activity makes it worse. They also suffer from disability in that common tasks like taking a shower or cooking dinner become difficult.

There is a huge cross-over between ME-CFS and musculoskeletal and spine conditions. Meaning that it's not uncommon for us to see chronic pain patients with neck or back issues who also carry an ME-CFS diagnosis. Since nobody knows what causes ME-CFS and there is no diagnostic test, the diagnosis is made based on symptoms of fatigue, which is also a common side effect of having chronic pain.

Many of our CCI patients also carry an ME/CFS diagnosis. The presumption is that their ME/CFS symptoms are caused by or made worse by the CCI.

MCAS/POTS

Mast cell activation syndrome (MCAS) and Postural Orthostatic Tachycardia Syndrome are overlapping diagnosis with CCI, mostly with patients who have EDS (23). MCAS involves overreactions of the immune system and unexplained allergic reactions and POTS is a fast heart rate when changing positions such as getting up from a chair or bed. Both of these diagnoses are quite common in our CCI patients with EDS.

It's believed that in CCI patients, POTS is caused by intermittent compression of the vagus nerve at C1/C2. This nerve is responsible for keeping your blood pressure high enough while you move to maintain your blood pressure. If the vagus nerve gets irritated, that can lead to a rapid heart rate and less blood pressure, leading to POTS symptoms.

Can MCAS cause ligament damage ? Here I only found a single publication linking mast cells to tendon damage (34). This paper theorizes that the neurologic inflammation that can be triggered by mast cells could theoretically damage tendons. However, it draws no direct link between the two. This is again, tendon damage and NOT ligament damage.

Dural Leaks

Most patients are unaware that there's fluid around their brain, spinal cord, and nerve roots that literally floats their whole neurologic system. They also don't realize that the system designed to hold it all can spring a leak and that when it does, all heck can break loose. While we've treated many patients with dural leaks through the years, one in particular stands out as a happenstance of being in the right place at the right time—a 16-year-old named Harry. His case is a great way to shed some light on dural leak treatment.

What Is the Dura?

Your nervous system is made up of your brain, the spinal cord, and the spinal nerves that leave the spine. There are also peripheral nerves that go to and from various parts of the body and a few other parts and pieces, but our focus today is on the central nervous system. To protect these fragile structures, your brain and spinal cord live inside strong bones (the skull and vertebrae).

Your body has a problem, and its solution dictates why dural leaks happen. The nerves and brain are sensitive structures that can be easily damaged. In addition, if you put any pressure on the nerves, they will fire off, giving a false signal, like a wire that's short-circuiting. To get around this, nature has enclosed all of the nerves into a sheath called the dura. It's kind of like a specialized water balloon that allows your brain, spinal cord, and nerves to float in liquid (called cerebrospinal fluid, or CSF). This provides excellent protection as well as shock absorption.

What Is a Dural Leak?

As you might imagine, at the end of the day, no system of containment is perfect. If you place enough pressure on even the world's best water balloon it can pop or spring a leak. In addition, if you poke a hole in it or if it was manufactured with a weak spot, the same thing can happen. The body is the same.

The containment system of the CSF, called the dura, can also spring a leak. When it does, the fluid can leak out and the brain, spinal cord, and nerves can lose their containment system.

What's unique about your personal water-balloon nerve-flotation system is that it's constantly topping off the system. To do this, your body makes CSF at a certain rate. If you spring a small leak, the body can handle that by simply ramping up the production of the fluid. However, if the volume per unit time of the leak exceeds the body's ability to replace the fluid, then you get "low in the tank," causing your brain to literally bang into the skull—talk about a headache!

What Are Dural Leak Symptoms?

Imagine an ice cream headache on steroids and multiply that by 20—that's what many patients with dural leaks experience. In addition, they can have a bevy of other nerve-type complaints because their spinal cord and nerves are being stretched. This can include confusion, disorientation, numbness or tingling, or pain referring to strange places. Given the vague nature of the complaints, it's not hard to see why these patients get lost in the medical care system. However, there is one symptom that's pretty specific to dural leak patients. They're better when lying flat, and it all gets worse when they're upright.

What Causes Dural Leaks?

The biggest cause is iatrogenic, which is a fancy medical way of saying that we doctors are the culprit. Some happen as a result of epidural injections that inadvertently puncture the dura. Some happen because of spinal taps, where the goal is to puncture the dura. Some can also occur in car crashes where sudden pressure builds up in the CSF, causing a dural blowout. Finally, some just happen (spontaneous), likely due to a pre-existing defect in the dura (a weak spot).

What Is a Dural Leak Treatment?

As silly as it sounds, the treatment for dural leak is as simple as "patch the hole"! Sometimes patients get offered a surgical repair, but that can be difficult if there are many holes to patch. In addition, finding where the leak is occurring in the first place can be maddeningly difficult as few tests exist to accurately show where the leak is located.

The old reliable dural leak treatment is called a blood patch. This is what it sounds like—injecting the patient's whole blood into the area around the dura (epidural) and allowing this to coagulate to seal the hole. The advantage of a blood patch is that you can inject several epidural sites and cover a whole area at a time, like the lumbar (low back) spine. If that doesn't work, you can march up to the lower thoracic, then upper thoracic, and then cervical. By process of elimination, you can thus find the region of the leak.

Some chronic dural leakers are also injected with fibrin glue, which is a newer dural leak treatment. This is the same stuff used to close skin wounds in the emergency room. It can be easily injected, and then it sets up with a rubber-cement-like consistency. We've heard mixed results from our dural leaker patients as some report that it can cause scarring of the dura, and at least one poor woman got hepatitis from a poorly screened human donor of the fibrin.

We've been blessed in this area with a natural dural leak treatment for many of these patients—platelet lysate (PL). We routinely inject epidural growth factors isolated from blood platelets to help patients with things like herniated discs. However, that same injectate turns out to be a great dural leak treatment as it can be easily polymerized to form what looks like a blood clot by injecting a little activation agent right after the PL. It also has a natural growth-factor cocktail to assist in healing the hole in the dura.

Retroflexed Dens

Here the dens is canted backward relative to the atlas. This can reduce the amount of room between the dens and the upper spinal cord and brainstem. Normally, people are born with a retroflexed dens and it doesn't cause problems. However, it can predispose someone to have more cervicomedullary syndrome when there is also laxity in key ligaments like the transverse. In this case, the dens can move backward

with flexion of the head and neck, causing more compression of the neural structures than would otherwise happen if the shape of the dens was normal.

COVID

Some CCI patients feel that their symptoms started after being diagnosed with COVID-19. Or at some later date after a COVID diagnosis, in a subset of these patients, CCI is diagnosed. Hence, it's natural to wonder if there is a connection between these two. Right now, we have no credible data that suggests that COVID-19 causes CCI or ligament injury in general.

So why would some patients report CCI symptoms after COVID? First, for an infection to be associated with a diagnosis, one must come on the heels of the other. Meaning that we would expect to see CCI symptoms beginning within days to weeks of a COVID-19 diagnosis. In these cases, it's more likely that the pro-inflammatory nature of having any infection (including COVID-19) likely brought out a latent and smoldering problem. Meaning that the CCI was likely there, but asymptomatic.

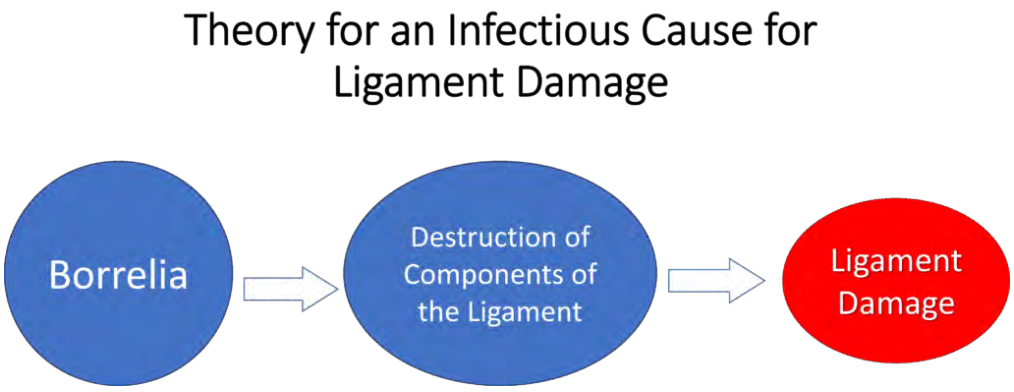
Another common association we see is that some patients feel that their symptoms began after getting a COVID-19 vaccine. Again, at this time, we have no mechanism that connects a vaccine and ligament injury or CCI. This again, is likely the same pro-inflammatory mechanism as described above.

Lyme Disease

The Basic Concepts

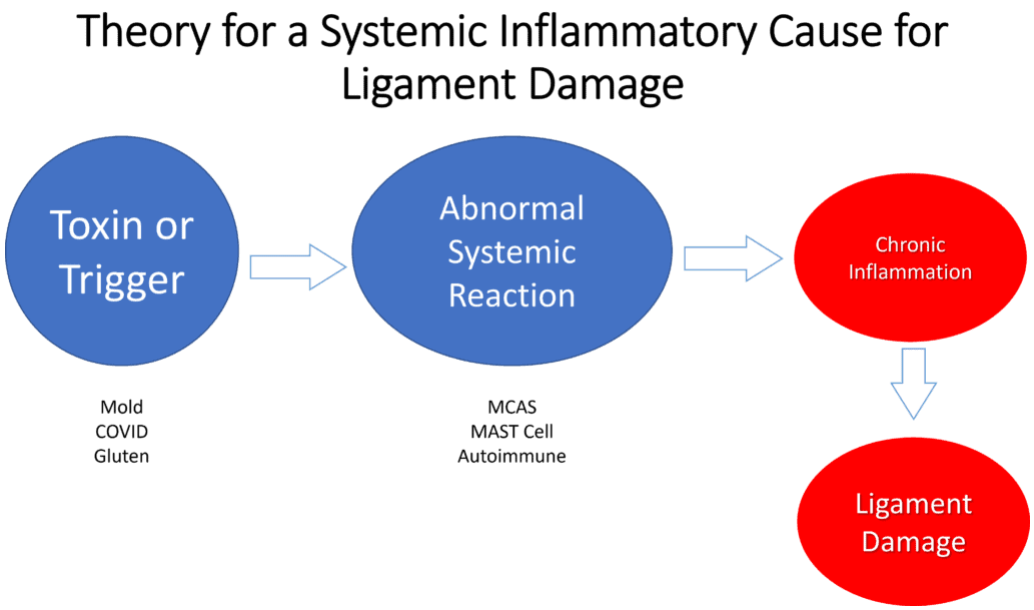
The two ideas we'll be dealing with today are ligament damage due to a pathogen eating away at connective tissue and damaging inflammation.

This is what the first theory looks like:



Here an infectious pathogen like Borrelia destroys ligaments by eating away at components of that structure.

The second theory we'll be covering today looks like this:



Here, some environmental or dietary trigger causes an abnormal immune system to respond with excessive inflammation which leads to ligament damage.

Remember, both of these are THEORIES without definitive scientific proof at this time.

Chronic Lyme Disease and Ligaments

The idea behind chronic Lyme disease is that a tick bite infects the patient with one of four species of the Borrelia spirochete. Acute and sub-acute Lyme infection can cause all sorts of issues from rashes to joint pain to neurologic symptoms. The more controversial issue is whether a long-term or chronic Lyme infection exists and whether that can cause widespread lifetime damage throughout the body. However, the task at hand here this morning is to determine if we have research connecting these Borrelia infections to ligament damage.

I searched the US National Library of Medicine for basic science studies for “Borrelia Ligament”. In published opinion/review pieces, I dove into the references in those publications and included any that seemed to show a credible link. This is what I found:

- A European dog study looked at Cranial Cruciate Ligament rupture (CCL) and Borrelia (and other vector-borne pathogens). No relationship was found between these pathogens and ligament rupture. (30) A lower-level canine case series did find a relationship between Borrelia and CCL rupture (33).
- A single case study of a patient who had Lyme spirochetes cultured from her ligament samples taken during a trigger finger surgery. (31)
- A case series report in German that connected C1-C2 instability (unknown if this was CCI or how that diagnosis was rendered) in some percentage of the cases examined who had chronic IgG antibodies to Borrelia and evidence of white blood cell sensitization to the same (LTT). (32) Functional MRIs were performed, but the exact data from those images was not really reported.

BOTTOM LINE: As far as convincing human research, there is one case series (lower-level evidence) of patients who had been diagnosed with CCI, and various tests for the body's response to Borrelia were positive, but this did not include a control group so we don't know how positive these tests would have been in random people from the local population. In addition, how CCI was diagnosed in this study is unknown. So, is there a clear connection between chronic Lyme and ligament damage? There's one study that looks interesting, but this is not in any way definitive.

Is it clear that Lyme disease causes CCI? No, there is no clear evidence that these two are causally associated.

Mold/CIRS

The general concept behind mold exposure is that the patient lives in a home where water damage leads to mold growth and that patient has the right genetic and environmental makeup to react to this exposure by producing a chronic inflammatory state. This syndrome goes by the name Chronic Inflammatory Response Syndrome (CIRS). I again did a search through the US National library of medicine, but regrettably found nothing linking mold or CIRS to ligament damage. In short, there is no credible connection between CIRS and CCI.

Chapter 9-Wrapping It All Up

Thanks for taking the time to dig into CCI. My goal here was to create a book that was simple enough to read that allowed patients to learn but also had enough information that physicians would learn new things as well. If you find any typos or feel that there are things missing that really need to be in the book, send me an email at centenooffice@centenoschultz.com. While I want to keep this book short enough to be a quick read, if 3-4 patients all request a section that's missing, I'll likely add that topic.

References

(1) Grabb PA1, Mapstone TB, Oakes WJ. Ventral brain stem compression in pediatric and young adult patients with Chiari I malformations. Neurosurgery. 1999 Mar;44(3):520-7; discussion 527-8. https://www.ncbi.nlm.nih.gov/pubmed/10069589

(2) Rojas CA, Bertozzi JC, Martinez CR, Whitlow J. Reassessment of the craniocervical junction: normal values on CT. AJNR Am J Neuroradiol. 2007 Oct;28(9):1819-23. doi: 10.3174/ajnr.A0660. Epub 2007 Sep 24. PMID: 17893223.

(3) Henderson FC Sr, Henderson FC Jr, Wilson WA 4th, Mark AS, Koby M. Utility of the clivo-axial angle in assessing brainstem deformity: pilot study and literature review. Neurosurg Rev. 2018;41(1):149–163.
doi:10.1007/s10143-017-0830-3

(4) Khanna G, Sato Y. Imaging of the craniovertebral junction. Oper Tech Neurosurg. 2005;8(3):131–142. doi: 10.1053/j.otns.2005.10.004.

(5) Tedeschi, Helder; Ghizoni, Enrico; Mathias, Roger N.; Fernandes, Yvens B.; Joaquim, Andrei F.; Batista, Ulysses C. (Apr 1, 2015). "Computed tomography evaluation of the normal craniocervical junction craniometry in 100 asymptomatic patients". Neurosurgical Focus. 38 (4): E5. doi:10.3171/2015.1.FOCUS14642. ISSN 1092-0684.

(6) Krakenes J1, Kaale BR. Magnetic resonance imaging assessment of craniovertebral ligaments and membranes after whiplash trauma. Spine (Phila Pa 1976). 2006 Nov 15;31(24):2820-6. PMID:17108836

(7) Dvorak J, Penning L, Hayek J, Panjabi MM, Grob D, Zehnder R. Functional diagnostics of the cervical spine using computer tomography. Neuroradiology. 1988;30(2):132-7. doi: 10.1007/BF00395614. PMID: 3386806.

(8) Gluf, W. M., & Brockmeyer, D. L. (2005). Atlantoaxial transarticular screw fixation: a review of surgical indications, fusion rate, complications, and lessons learned in 67 pediatric patients, Journal of Neurosurgery: Spine, 2(2), 164-169.

(9) Ghostine SS, Kaloostian PE, Ordookhanian C, et al. Improving C1-C2 Complex Fusion Rates: An Alternate Approach. Cureus. 2017;9(11):e1887. Published 2017 Nov 29. doi: 10.7759/cureus.1887

(10) Schroeder GD, Hsu WK. Vertebral artery injuries in cervical spine surgery. Surg Neurol Int. 2013;4(Suppl 5):S362–S367. Published 2013 Oct 29. doi:10.4103/2152-7806.120777

(11) Myers KD, Lindley EM, Burger EL, Patel VV. C1-C2 fusion: postoperative C2 nerve impingement-is it a problem?. Evid Based Spine Care J. 2012;3(1):53–56. doi: 10.1055/s-0031-1298601

(12) Rybarczyk M, Baranowska A, Baranowski P. Assessment of the results of occipito-cervical stabilization in cranio-vertebral damage. Pol Merkur Lekarski. 2020 Aug 22;49(286):228-231. PMID: 32827415.

(13) De Vlieger J, Dejaegher J, Van Calenbergh F. Posterior fossa decompression for Chiari malformation type I: clinical and radiological presentation, outcome and complications in a retrospective series of 105 procedures. Acta Neurol Belg. 2019 Jun;119(2):245-252. doi: 10.1007/s13760-019-01086-7. Epub 2019 Feb 8. PMID: 30737652.

(14) Furuya K, Sano K, Segawa H, Ide K, Yoneyama H. Symptomatic tonsillar ectopia. J Neurol Neurosurg Psychiatry. 1998;64(2):221–226. doi:10.1136/jnnp.64.2.221

(15) Freeman MD, Rosa S, Harshfield D, Smith F, Bennett R, Centeno CJ, Kornel E, Nystrom A, Heffez D, Kohles SS. A case-control study of cerebellar tonsillar ectopia (Chiari) and head/neck trauma (whiplash). Brain Inj. 2010;24(7-8):988-94. doi: 10.3109/02699052.2010.490512.

(16) Dubey A, Sung WS, Shaya M, Patwardhan R, Willis B, Smith D, Nanda A. Complications of posterior cranial fossa surgery–an institutional experience of 500 patients. Surg Neurol. 2009 Oct;72(4):369-75. doi: 10.1016/j.surneu.2009.04.001. Epub 2009 Jul 14.

(17) Düz B, Gocmen S, Secer HI, Basal S, Gönül E. Tethered cord syndrome in adulthood. J Spinal Cord Med. 2008;31(3):272-278. doi:10.1080/10790268.2008.11760722

(18) Talamonti G, Marcati E, Mastino L, Meccariello G, Picano M, D'Aliberti G. Surgical management of Chiari malformation type II. Childs Nerv Syst. 2020 Aug;36(8):1621-1634. doi: 10.1007/s00381-02004675-7. Epub 2020 May 30. PMID: 32474814.

(19) Mancarella C, Delfini R, Landi A. Chiari Malformations. Acta Neurochir Suppl. 2019;125:89-95. doi: 10.1007/978-3-319-62515-7_13. PMID: 30610307.

(20) Garcés-Ambrossi GL, McGirt MJ, Samuels R, Sciubba DM, Bydon A, Gokaslan ZL, Jallo GI. Neurological outcome after surgical management of adult tethered cord syndrome. J Neurosurg Spine. 2009 Sep;11(3):304-9. doi: 10.3171/2009.4.SPINE08265. PMID: 19769511.

(21) Shih P, Halpin RJ, Ganju A, Liu JC, Koski TR. Management of recurrent adult tethered cord syndrome. Neurosurg Focus. 2010 Jul;29(1):E5. doi: 10.3171/2010.3.FOCUS1073. PMID: 20594003.

(22) Garg K, Tandon V, Kumar R, Sharma BS, Mahapatra AK. Management of adult tethered cord syndrome: our experience and review of literature. Neurol India. 2014 Mar-Apr;62(2):137-43. doi: 10.4103/0028-3886.132329. PMID: 24823721.

(23) Kohn A, Chang C. The Relationship Between Hypermobile Ehlers-Danlos Syndrome (hEDS), Postural Orthostatic Tachycardia Syndrome (POTS), and Mast Cell Activation Syndrome (MCAS). Clin Rev Allergy Immunol. 2020 Jun;58(3):273-297. doi: 10.1007/s12016-019-08755-8. PMID: 31267471.

(24) Bogduk N, Govind J. Cervicogenic headache: an assessment of the evidence on clinical diagnosis, invasive tests, and treatment. Lancet Neurol. 2009 Oct;8(10):959-68. doi: 10.1016/S1474-4422(09)70209-1. PMID: 19747657.

(25) Panjabi M, Dvorak J, Crisco JJ 3rd, Oda T, Wang P, Grob D. Effects of alar ligament transection on upper cervical spine rotation. J Orthop Res. 1991 Jul;9(4):584-93. doi: 10.1002/jor.1100090415. PMID: 2045985.

(26) Nolet, P.S.; Nordhoff, L.; Kristman, V.L.; Croft, A.C.; Zeegers, M.P.; Freeman, M.D. Is Acceleration a Valid Proxy for Injury Risk in Minimal Damage Traffic Crashes? A Comparative Review of Volunteer, ADL and Real-World Studies. Int. J. Environ. Res. Public Health 2021, 18, 2901. https://doi.org/10.339ijerph18062901

(27)Henderson FC Sr, Henderson FC Jr, Wilson WA 4th, Mark AS, Koby M. Utility of the clivo-axial angle in assessing brainstem deformity: pilot study and literature review. Neurosurg Rev. 2018 Jan;41(1):149-163. doi: 10.1007/s10143-017-0830-3. Epub 2017 Mar 3. PMID: 28258417; PMCID: PMC5748419.

(28) Nicholson LL, Rao PJ, Lee M, Wong TM, Cheng RHY, Chan C. Reference values of four measures of craniocervical stability using upright dynamic magnetic resonance imaging. Radiol

Med. 2023 Mar;128(3):330-339. doi: 10.1007/s11547-023-01588-8. Epub 2023 Jan 30. PMID: 36715785; PMCID: PMC10020271.

(29) Bai C, Wang Z, Guan J, et al. Clinical characteristics and neuroimaging findings in eagle syndrome induced internal jugular vein stenosis. Ann Transl Med. 2020;8(4):97. doi:10.21037/atm.2019.12.93

(30) Tabar MD, Tabar J, Naranjo C, Altet L, Roura X. Detection of vector-borne pathogens in owned dogs with cranial cruciate ligament rupture living in the Mediterranean area. Parasit Vectors. 2022 May 10;15(1):105. doi: 10.1186/s13071-022-05205-x. PMID: 35534857; PMCID: PMC9088045.

(31) Häupl T, Hahn G, Rittig M, Krause A, Schoerner C, Schönherr U, Kalden JR, Burmester GR. Persistence of Borrelia burgdorferi in ligamentous tissue from a patient with chronic Lyme borreliosis. Arthritis Rheum. 1993 Nov;36(11):1621-6. doi: 10.1002/art.1780361118. PMID: 8240439.

(32) Müller KE. Erkrankungen der elastischen und kollagenen Fasern von Haut, Sehnen und Bänder bei Lyme-Borreliose. umwelt-medizin-gesellschaft. 2009;22(2):112–8.

(33) Muir P, Oldenhoff WE, Hudson AP, Manley PA, Schaefer SL, Markel MD, Hao Z. Detection of DNA from a range of bacterial species in the knee joints of dogs with inflammatory knee arthritis and associated degenerative anterior cruciate ligament rupture. Microb Pathog. 2007 Feb-Mar;42(2-3):47-55. doi: 10.1016/j.micpath.2006.10.002. Epub 2007 Feb 21. PMID: 17320342.

(34) Alim MA, Peterson M, Pejler G. Do Mast Cells Have a Role in Tendon Healing and Inflammation? Cells. 2020 May 4;9(5):1134. doi: 10.3390/cells9051134. PMID: 32375419; PMCID: PMC7290807.

(35) Berger DR, Centeno CJ, Kisiday JD, McIlwraith CW, Steinmetz NJ. Colony Forming Potential and Protein Composition of Commercial Umbilical Cord Allograft Products in Comparison With Autologous Orthobiologics. Am J Sports Med. 2021 Oct;49(12):3404-3413. doi: 10.1177/03635465211031275. Epub 2021 Aug 16. PMID: 34398643.

(36) Osmotherly PG, Farrell SF, Digby SD, Rowe LJ, Buxton AJ. The influence of age, sex, and posture on the measurement of atlantodental interval in a normal population. J Manipulative Physiol Ther. 2013 May;36(4):226-31. doi: 10.1016/j.jmpt.2013.04.004. Epub 2013 May 8. PMID: 23664159.