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The Craniocervical Junction: Observations regarding the Relationship between Misalignment, Obstruction of Cerebrospinal Fluid Flow, Cerebellar Tonsillar Ectopia, and Image-Guided Correction

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Abstract

The craniocervical junction is the most complex area of the spinal axis. Due to its complexity it is extremely vulnerable to injuries to the soft tissue stabilizing ligaments and membranous structures. Proper imaging in this area is an essential key to proper diagnosis directing towards the most appropriate and safe treatment options when injury occurs. Misalignments of C_0–C_1, C_1–C_2 brought on by head or neck trauma can manifest in different outcomes. Some of those outcomes can affect or cause neural compromise, and/or some may contribute to cerebrospinal fluid (CSF) flow obstruction as well as arteriovenous compromise. C_1 misalignment may also contribute to distention of the cerebellar tonsils (cerebellar tonsillar ectopia), i.e. down through the foramen magnum due to caudal tension by way of dentate ligament pathological stress on the spinal cord leading to obstruction of the normal flow of CSF. Mechanical compression of the jugular vein by the transverse process of C_1 has been found to lead to obstruction of outgoing venous blood flow. Such obstruction has been found in chronic cerebral spinal venous insufficiency which has been observed in neurodegenerative brain diseases such as multiple sclerosis. Image-Guided Atlas Treatment™ (IGAT™) has been shown to be a method of gentle correction of misalignment of C_0–C_1, C_1–C_2, resulting in improved CSF flow as well as venous outflow. Image-guided atlas treatment utilizes advanced dynamic upright MRI as the means of evaluating misalignments at the craniocervical junction, and the images obtained are used to calculate the appropriate alignment vectors to correct the misalignment. Post-correction advanced upright MRI images are then used to validate the appropriate realignment of C_0–C_1, C_1–C_2 to establish improvement in proper CSF as well as arteriovenous flow.

Understanding the anatomy and physiology of the craniocervical junction (CCJ) is essential in order to properly assess and correlate injuries to symptoms. The ability of the clinician to gather an accurate history of any traumatic event helps to determine the plausible insulting force vector in order to understand where forces and or stress might have led to the 'craniocervical syndrome'. It is important to correlate the patient's symptomatic picture to structures at the CCJ that might be eliciting their symptoms. The traditional intervertebral disc on cord or disc on exiting nerve root does not occur at the CCJ as there are no discs between C_0–C_1 and C_1–C_2. Looking for pathology that could radiate in or out from the CCJ must be correlated to the structures involved.

Anatomy/Physiology of the CCJ

It is important to consider that the central nervous system is encased in bone. While the literature describes our present understanding of the anatomy of the brain and spinal cord, the orientation of the spinal structures and the impact they have on central nervous system function is not as well understood [1, 2].

The location of the foramen magnum in relation to the center of mass of the skull as well as the shape, size and orientation of the skull and occipital condyles all impact on the weight-bearing characteristics of the cervical spine. The size, height and angle of the odontoid process and facet joints at each level of the spine have an impact on the contour of the cervical spine region and will affect the total size and shape of the lordotic curve.

The skull balances upon the atlas, with the occipital condyles resting on the superior facets. The atlas is attached to the axis, with the posterior surface of the anterior arch of the atlas being held to the facet on the anterior surface of the dens. The transverse cruciate ligament holds the atlas in place, allowing rotation while limiting anterior translational displacement of the atlas. The atlas also attaches to the axis with inferior facets resting on the superior facets of the axis. The absence of a vertebral body of the atlas and the large odontoid process of the axis make the two vertebrae of the CCJ unique.

The occiput/atlas/axis complex (CCJ) is stabilized by the following ligamentous and membranous structures. The alar ligaments usually attach the medial occipital condyles to the odontoid process. In a small percentage of cases, the alar ligaments attach to the anterior arch of the atlas. The apical ligament, tectorial membrane, anterior atlanto-occipital and atlantoaxial membranes stabilize the anterior structures of the CCJ. These structures, along with the posterior atlanto-occipital and atlantoaxial membranes, stabilize the flexion and extension of the occiput on atlas and the atlas on axis [3].

The vertebral arteries pass through the foramina of the transverse processes of the atlas. They make a 90° turn through the transverse foramina of C_1, and wrap around the posterior border of the lateral masses of C_1, making another 90° turn up into and through the foramen magnum to conjoin to become the basilar artery. The jugular veins pass just anterior to these transverse processes. The spinal cord passes through the neural canal of the atlas and axis as it traverses down the spinal canal. The role of the denticulate ligaments is to keep the cord centered in the neural canal and prevent the movements of the spine (flexion, extension, lateral flexion, etc.) from causing stress or tension on the cranial elements, and further to prevent the cord from impacting upon the anterior and posterior portion of the bony spinal canal [4].

Together, the shape and size of each of these CCJ structures contribute to the overall function and stability of the spine. When the balance of these structures is disturbed by trauma, there can be significant impairment as a result [5]. Abnormal tension on the cord can be caused by loss of the cervical lordosis or other misalignments of the spine. Tension on the cord can cause a caudal migration of the cerebellar tonsils known as cer-

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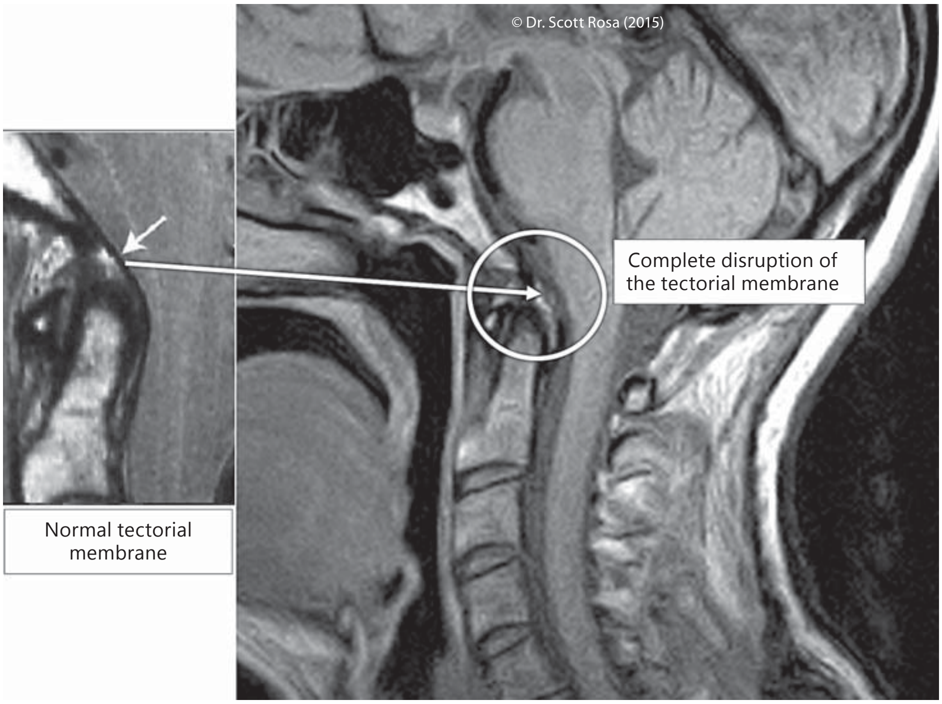

Fig. 1. Disruption of the tectorial membrane.

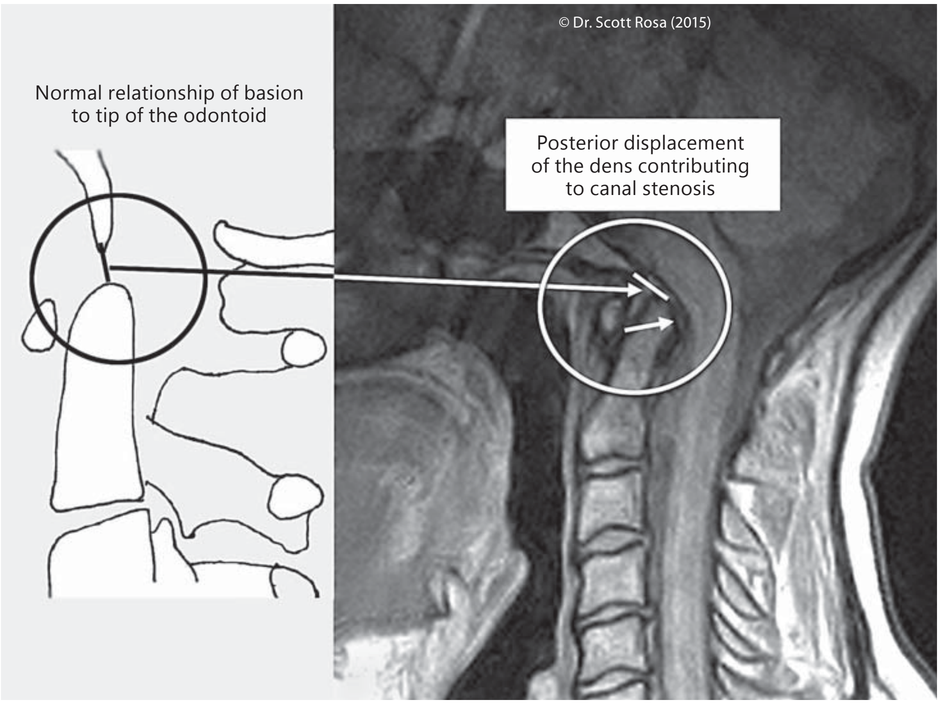

Fig. 2. Posterior migration of the dens.

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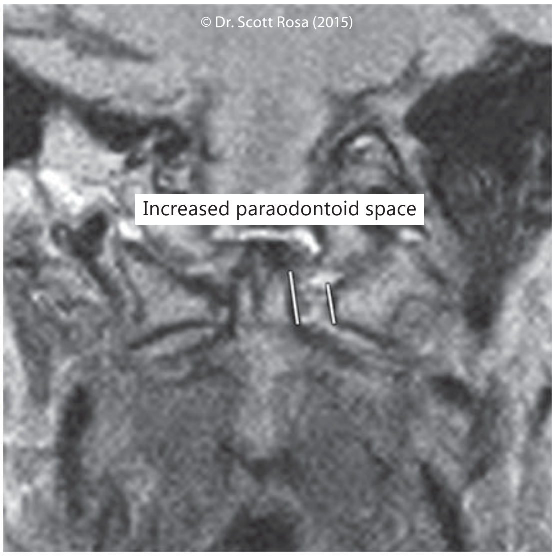

Fig. 3. Increased paraodontoid interspace.

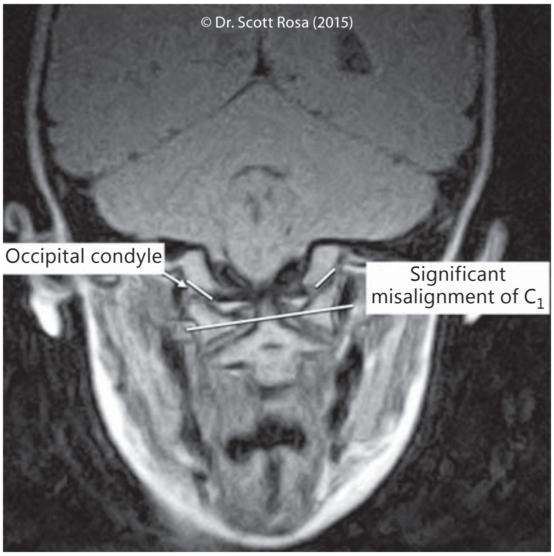

Fig. 4. Atlas/axis rotation on the x-axis.

ebellar tonsillar ectopia (CTE) [6]. Due to the gravitational effect of weight bearing, CTE is observed more frequently in upright MRI than in recumbent MRI [7]. In the recumbent position, the foramen magnum is oriented vertically and the cerebellum will have a gravitational force directing it away from the foramen magnum toward the posterior occiput. Since the upright MRI permits imaging of the CCJ in weight bearing in the same way stress radiography and videofluoroscopy or motion X-ray does, the effect of gravity allows visualization of the structural misalignment under the load of the skull mass [8, 9].

Abnormal flow of cerebrospinal fluid (CSF) can occur as a result of the cerebellar tonsils obstructing the foramen magnum, preventing CSF from properly circulating through and around the brain and down the spinal column to bring nutrients in and remove metabolic wastes from both the brain and spinal cord. Traumatic disturbance of spinal alignment can also impose abnormal tension on the vasculature, causing stretching or compression resulting in impaired arterial and/or venous flow. Abnormal loading can cause disruption of the other soft tissue, such as the facet capsular ligaments, which are capable of triggering nociceptive and proprioceptive disturbances in the ligaments and joint capsules, causing pain. These injuries have been found fairly often in patients with head/neck trauma, especially those involved in motor vehicle accidents.

Accident history provides important information about the force vector as well as significant risk factors which may result in more complicated injury. Risk factors in motor vehicle accidents would include: having the head turned during impact [10], rear-end impact, head restraint geometry, the relative size of vehicles in a collision, air bag deployment, preexisting spinal degeneration, awareness before impact and seat damage from impact [11], to name but a few.

Misalignments of the CCJ

Misalignments at the CCJ are a fairly common finding observed on plain film X-rays. The atlas can misalign in a 3D plane, including lateral dis-

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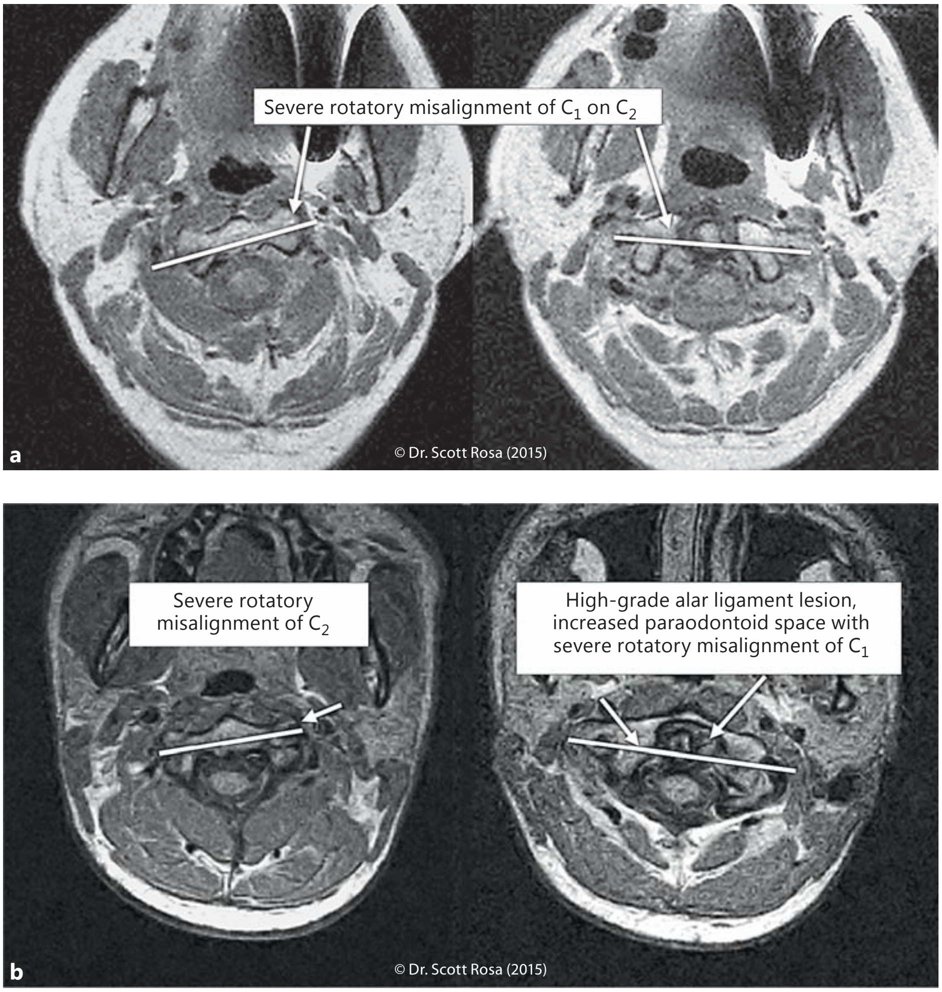

Fig. 5. a, **b** Atlas/axis rotation on the y-axis.

placement up and around the occipital condyle as well as rotational misalignment. Coupled with C_2 misalignments, especially counter rotation of C_1–C_2, this constitutes a problem as many physicians have not been trained to look for these pathognomonic findings. While fractures, listhesis and pathology such as tumors are consistently diagnosed, numerous conditions emanating from the CCJ are of clinical importance. While not as well understood, injuries at the CCJ can manifest in symptoms that are directly driven by soft tissue structures such as ligaments and membranous structures, which have nociceptive capabilities. It is essential that the practitioner understands that the CCJ is an area that is at increased risk of injury and instability due to its high degree of mobility. Careful observations with advanced imaging techniques demonstrate these misalignments on upright MRI. These misalignments can influence numerous pathophysiological conditions, including nerve compression, vascular compromise (vertebral artery insufficiency), as well as al-

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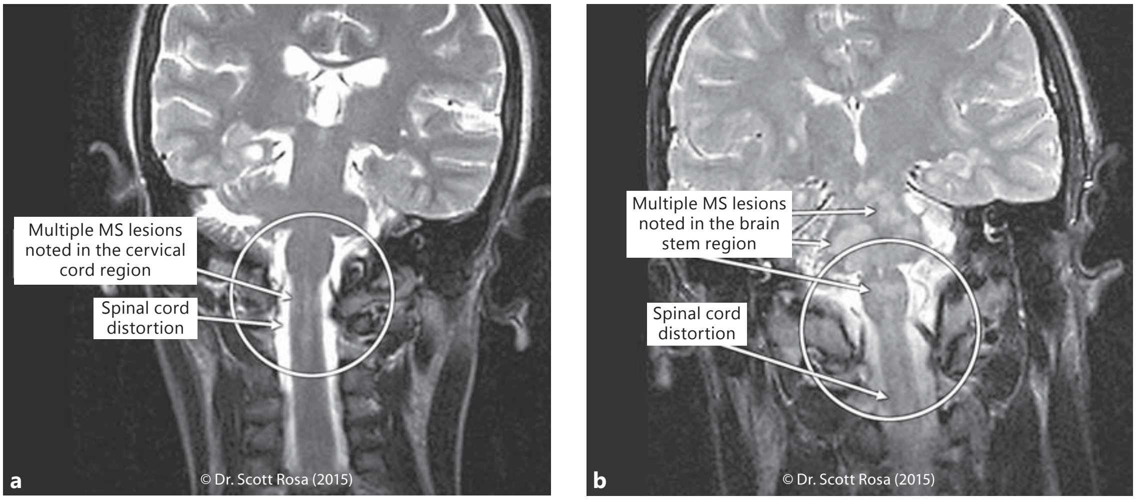

Fig. 6. a, **b** Spinal cord distortion.

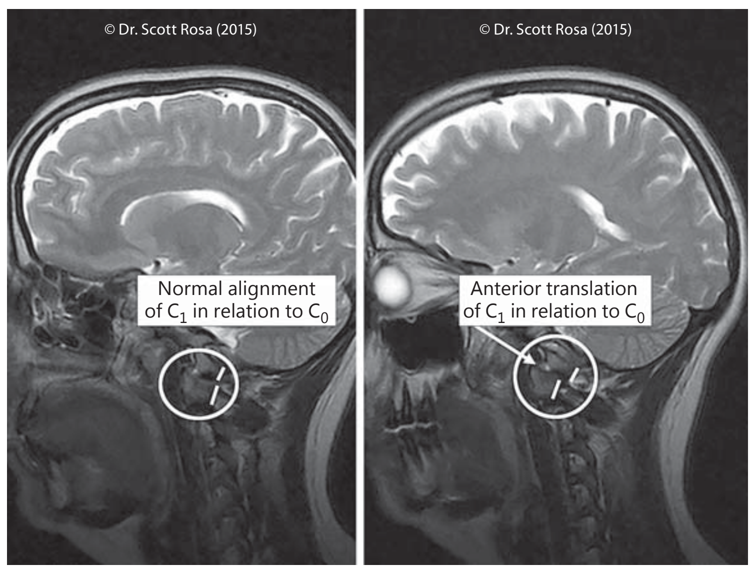

Fig. 7. Anterior/posterior translation of C_0/C_1.

tering the normal flow of spinal fluid in and out of the cranial vault. Misalignments at the CCJ of C_1–C_2 have also been implicated in the potential compression of the exiting jugular veins, which has been discussed in the literature. The effects of these misalignments can manifest in numerous ways. In some patients, CCJ misalignments can cause headaches, neck pain (skull base pain), nausea, dizziness, tinnitus and facial pain, to name but a few. Chronic misalignment can result in morphologic changes in the spinal musculature [12].

Fatty infiltration of the rectus capitis minor muscle has been observed in patients with chronic whiplash. Misalignment (C_0–C_1, C_1–C_2) at the CCJ can also influence and have an effect on the

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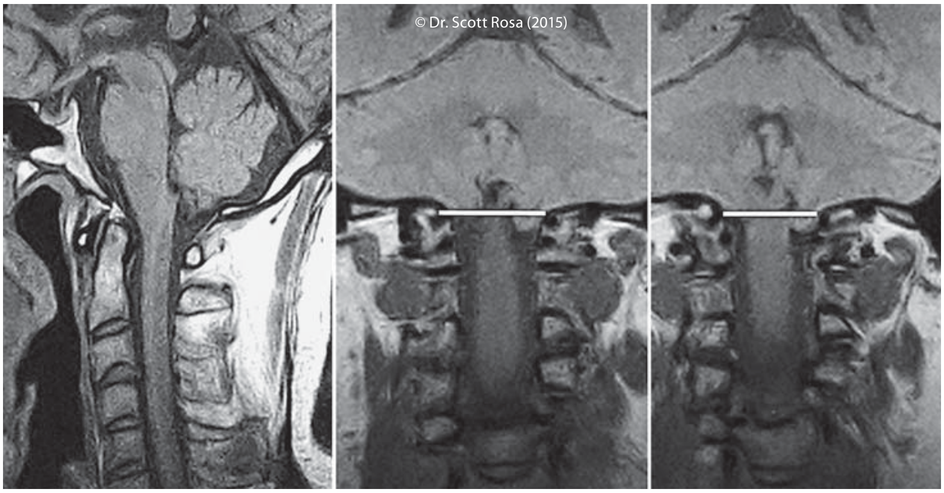

Fig. 8. Low cerebellar tonsils. Sagittal imaging alone is insufficient to assess CTE. Sequential coronal slices demonstrate a low-lying cerebellar tonsil on the left.

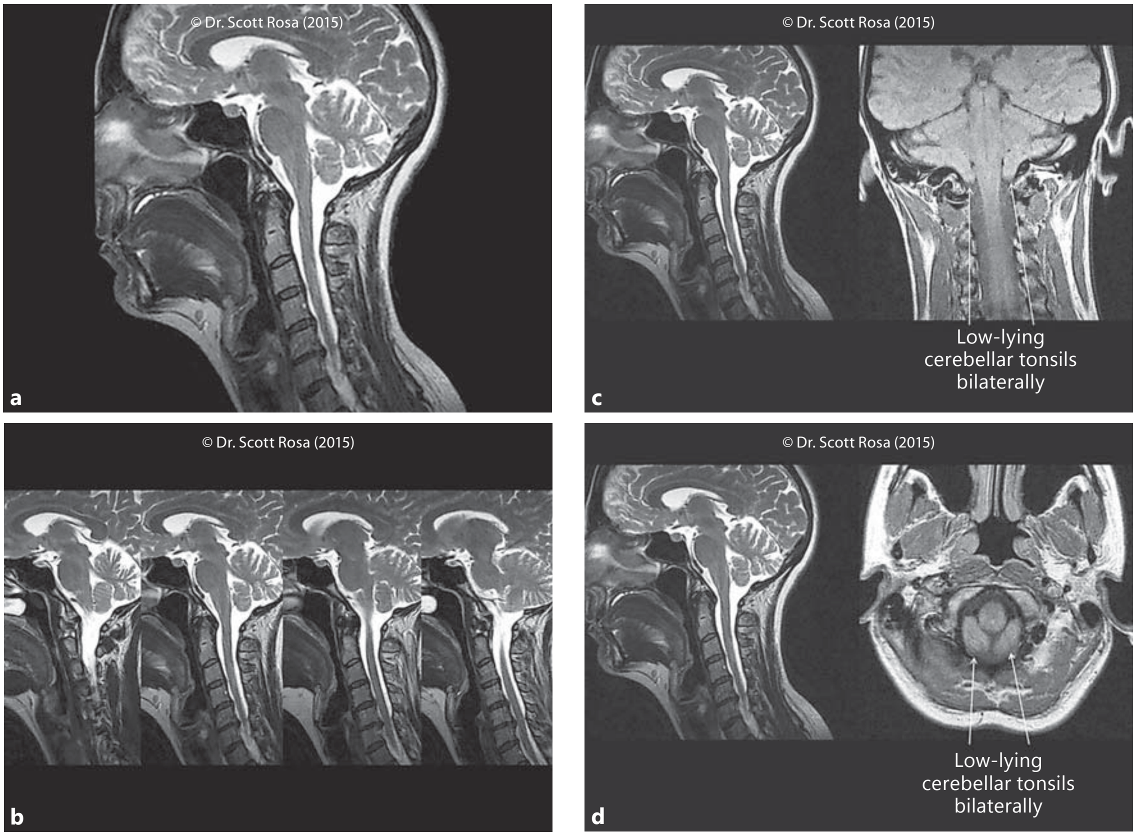

Fig. 9. Sagittal sequence denoting normal tonsillar position (**a**, **b**), whereas both coronal and axial images demonstrate significant CTE (**c**, **d**). Coronal and axial images are more sensitive and specific to identifying CTE and should be included in any imaging study of the craniocervical junction.

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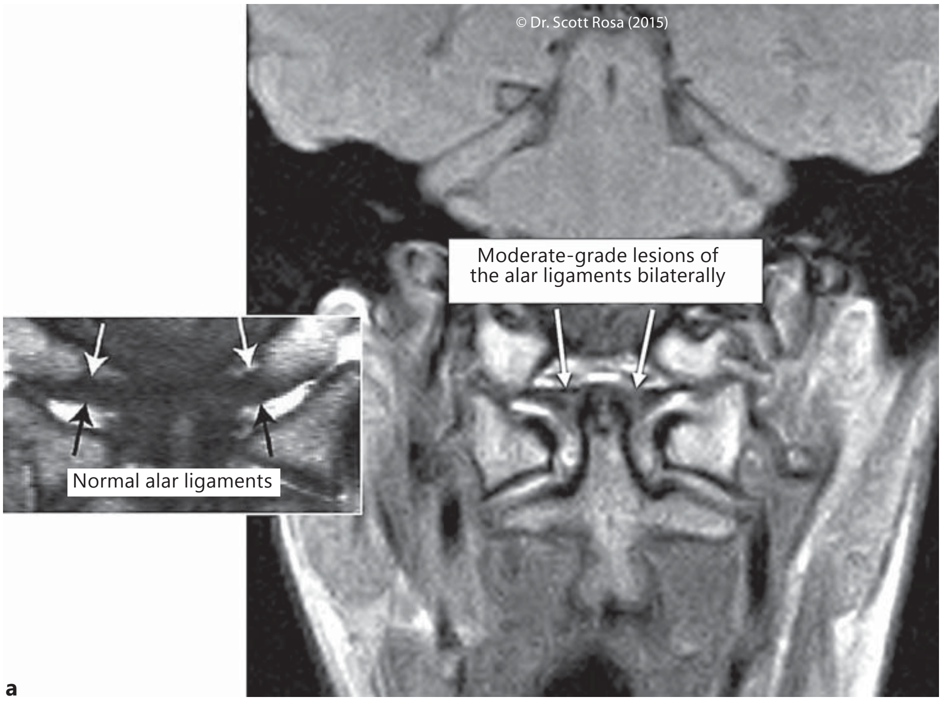
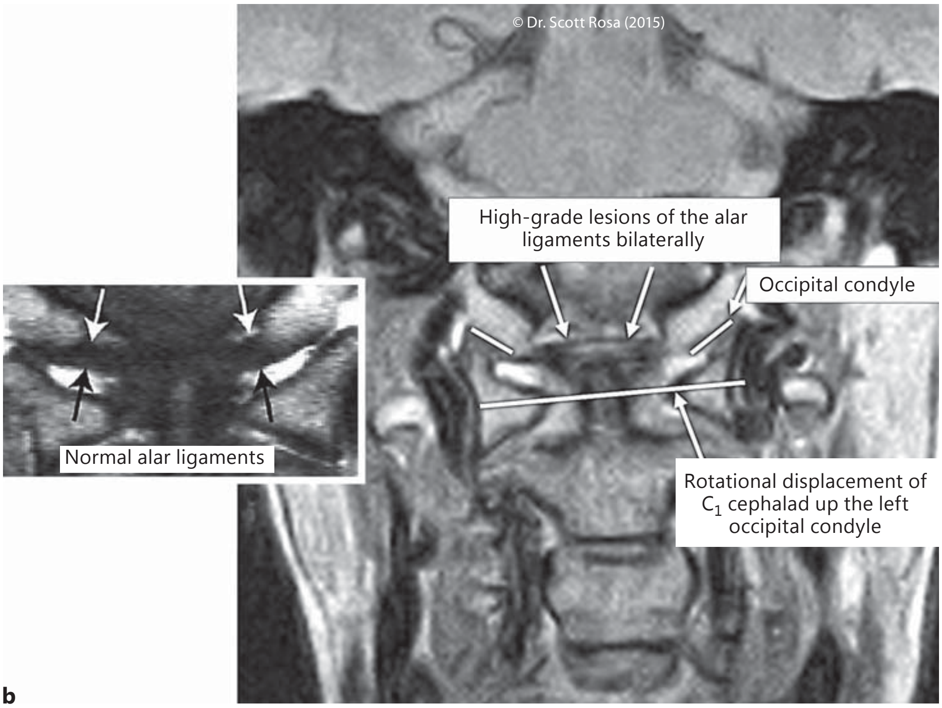


a



b

Fig. 10. a, **b** The alar ligaments.

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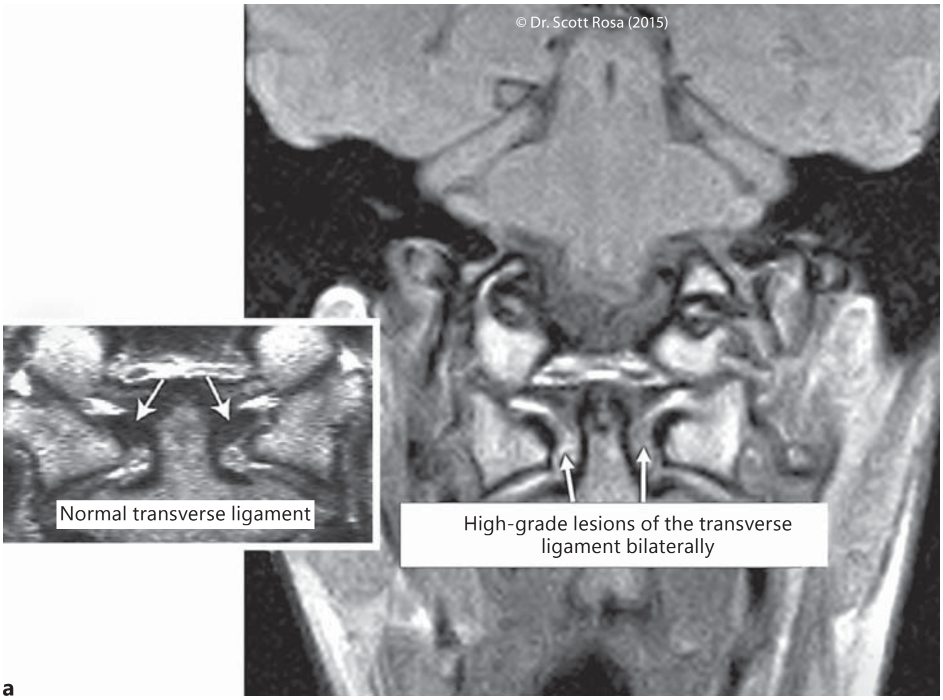
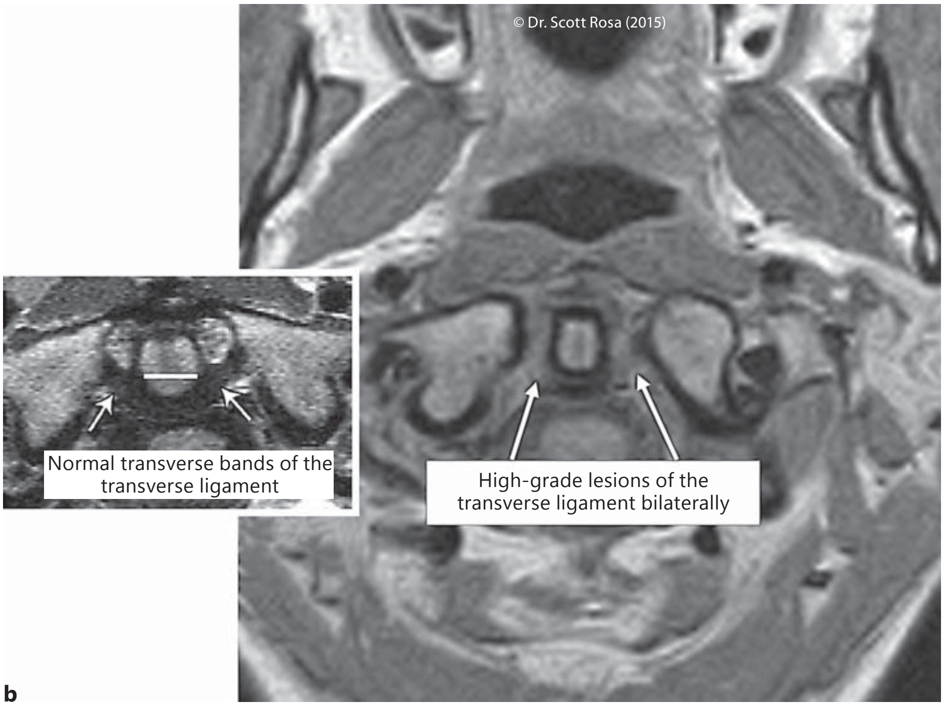


a



b

Fig. 11. a, **b** The transverse ligaments.

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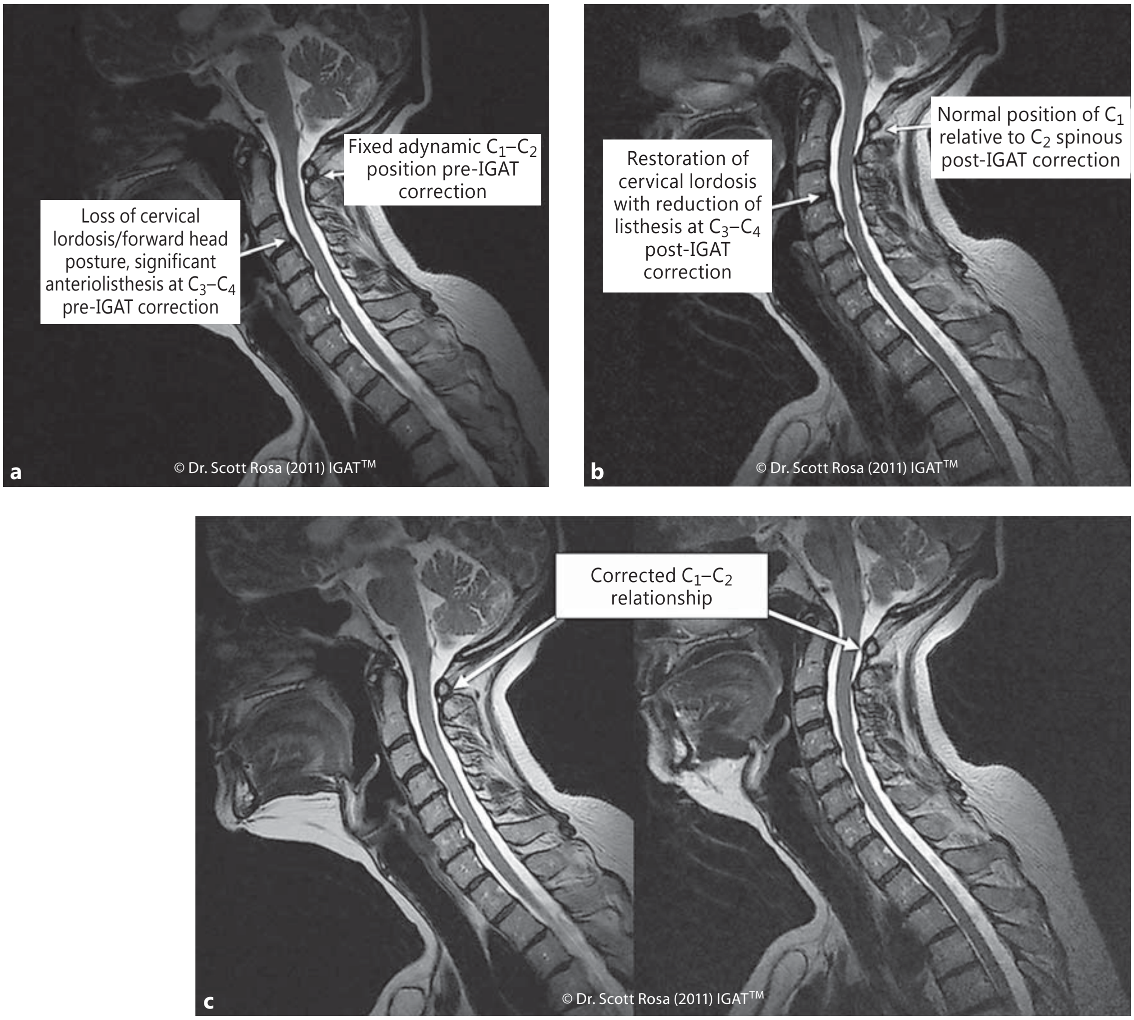

Fig. 12. Case 1: pre-IGAT correction (**a**); post-IGAT correction (**b**); corrected C_1–C_2 relationship (**c**).

cerebellar tonsillar position (due to the attachment of the dentate ligaments), which may impair or obstruct the normal flow of CSF. CSF obstruction may contribute to intracranial pressure (normal pressure hydrocephalus) and further manifest in more progressive symptoms and/or conditions. Altered CSF flow has been implicated in neurodegenerative brain diseases in the literature of late, including multiple sclerosis, Parkinson's disease and dementia, etc.

It has been postulated that misalignments of C_1–C_2 can contribute to cord distortion via the dentate ligaments. They can produce direct neurological insult via direct mechanical irritation, and indirectly via vascular compromise of the cervical cord. It has been further discussed that cord stress may produce venous occlusion with stasis of blood and resulting anoxia in particular areas of the upper cervical cord.

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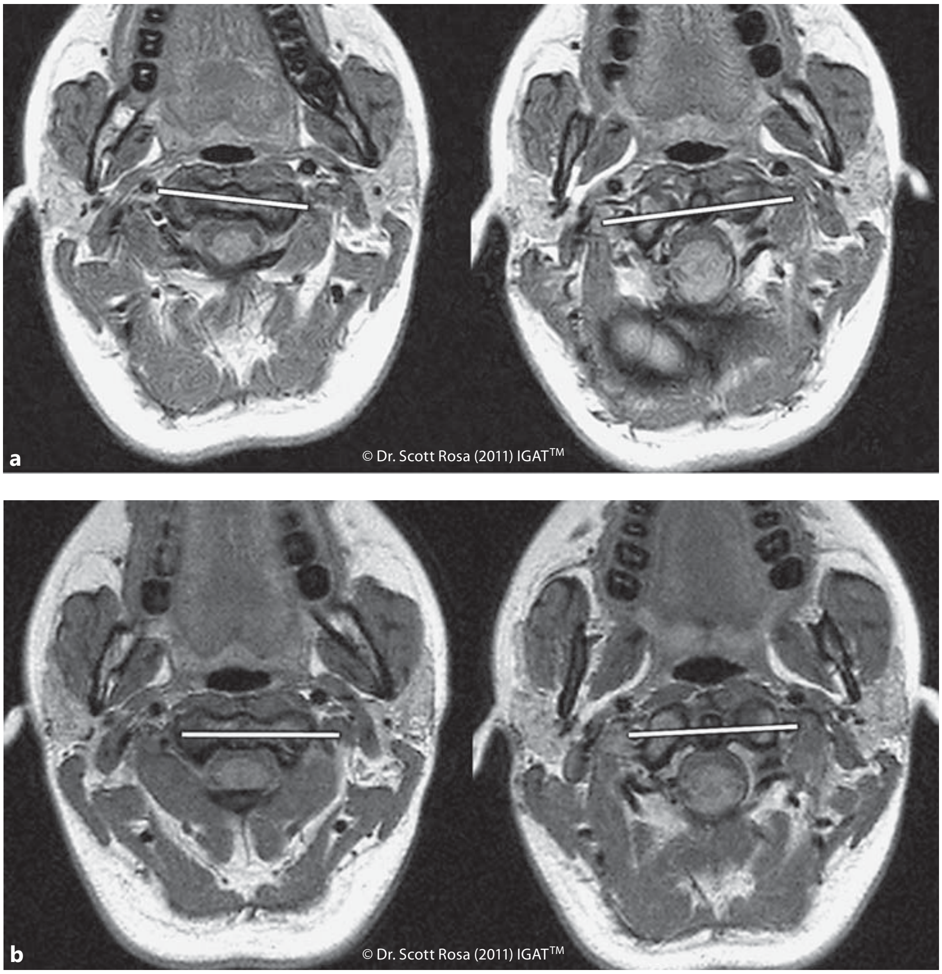

Fig. 13. Case 2: malrotation of C_1 in relation to C_2 pre-IGAT correction (**a**); substantial reduction in misalignment of C_1 in relation to C_2 post-IGAT correction (**b**).

Vertebral Artery Insufficiency

Misalignments of the CCJ have been shown to contribute to vertebral artery insufficiency. Vertebrobasilar insufficiency or vertebral basilar ischemia refers to temporary symptoms due to decreased blood flow in the posterior circulation of the brain. This may manifest as dizziness, but in more severe cases may manifest as weakness of the quadriceps muscles, causing a buckling of the knees where the legs give out in what is referred to as a 'drop attack'.

Cerebellar Tonsillar Ectopia

Misalignments of the CCJ have been found to influence the proper flow of CSF. Caudally displaced cerebellar tonsils may be due to a congenital malformation or may be acquired and have been observed with significant frequency in the whiplash injury population. Previously asymptomatic CTE due to congenital malformation (Chiari) may become symptomatic after head trauma [13]. Migration of the cerebellar tonsils

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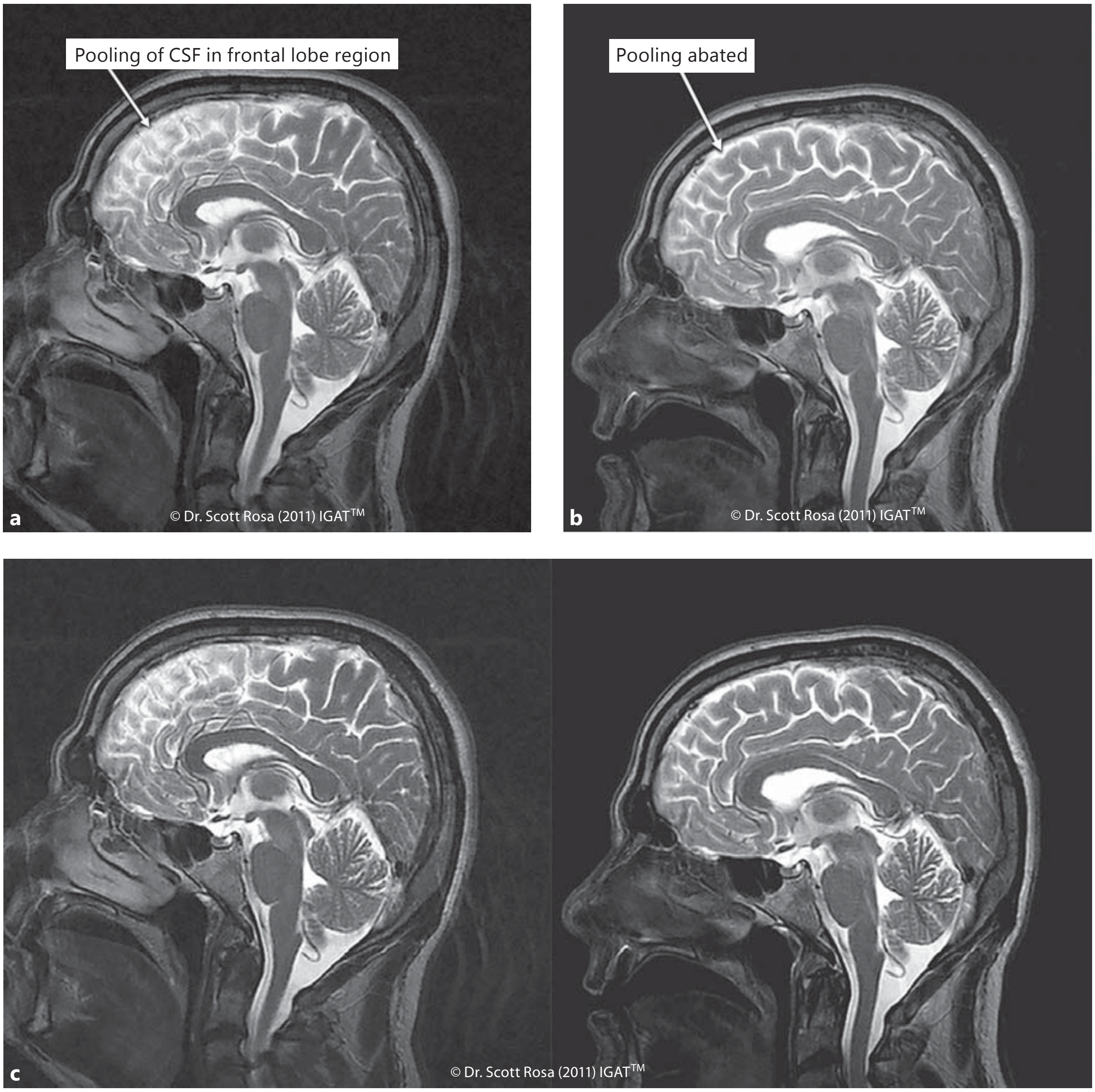

Fig. 14. Case 3: pre-IGAT C_1 correction (**a**); post-IGAT C_1 correction (**b**); **c** Pre-IGAT (left) and post-IGAT (right) C_1 correction.

may result in aberrant flow of CSF, which is observable on CSF flow studies [14–16]. Spinal cord tethering appears to be a plausible contributor of acquired CTE in cases where the C_1–C_2 misalignment is corrected and improvement in CSF flow has been demonstrated.

The dentate ligaments attach to the dural covering, affecting the behavior of the spinal cord tissue as forces generated in the dura are transmitted to the spinal cord tissue. When the dura is pulled taut with increasing canal length, tension is transmitted from the dentate ligaments to the cord. Tension in the dentate ligaments stabilizes the

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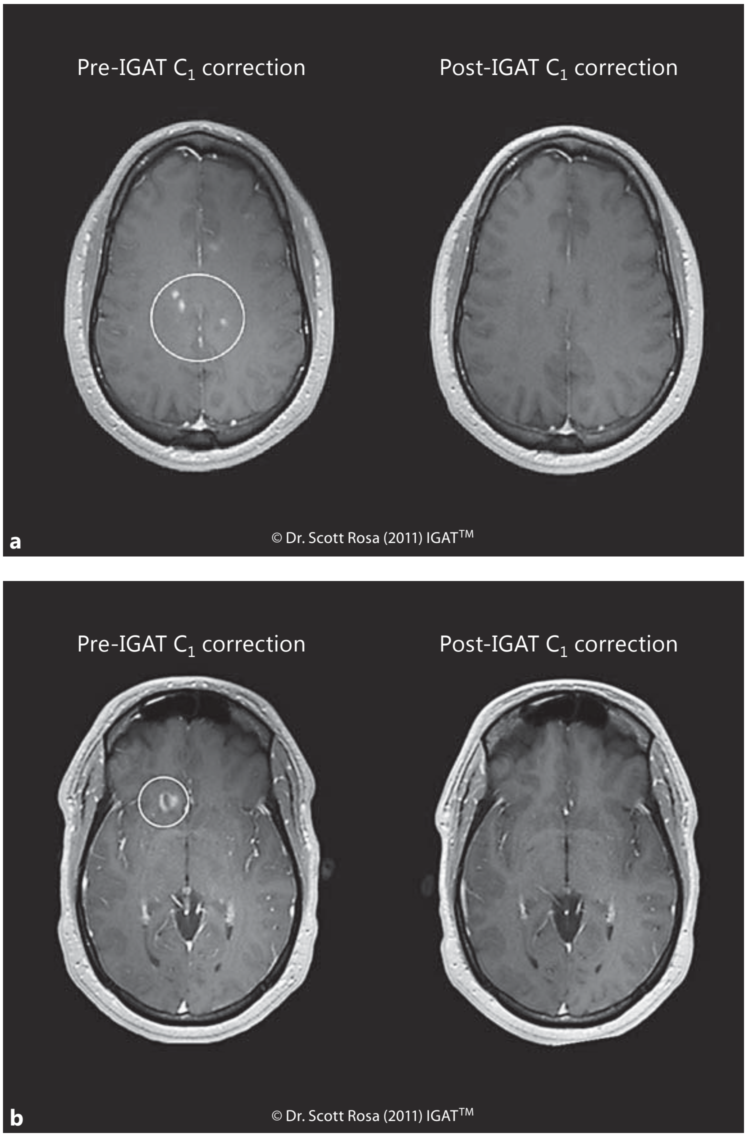

Fig. 15. a, **b** Case 4: sclerotic lesions in a patient diagnosed with multiple sclerosis which have abated after IGAT C_1 correction.

cord in the canal, but it can also have a tethering or overstretching effect on the cord. In cases where dentate ligament tension on the cord persists, morphological changes in the spinal cord tissue, such as the plaques seen in multiple sclerosis, may occur [17].

Diagnostic Imaging

Imaging of the CCJ has its limitations. Presently, the ability to properly image the CCJ in a 3D plane has been somewhat limited. Proper imaging of patients involved in a trauma is needed to rule out catastrophic injury (fracture, dislocation, loss of consciousness or possible internal

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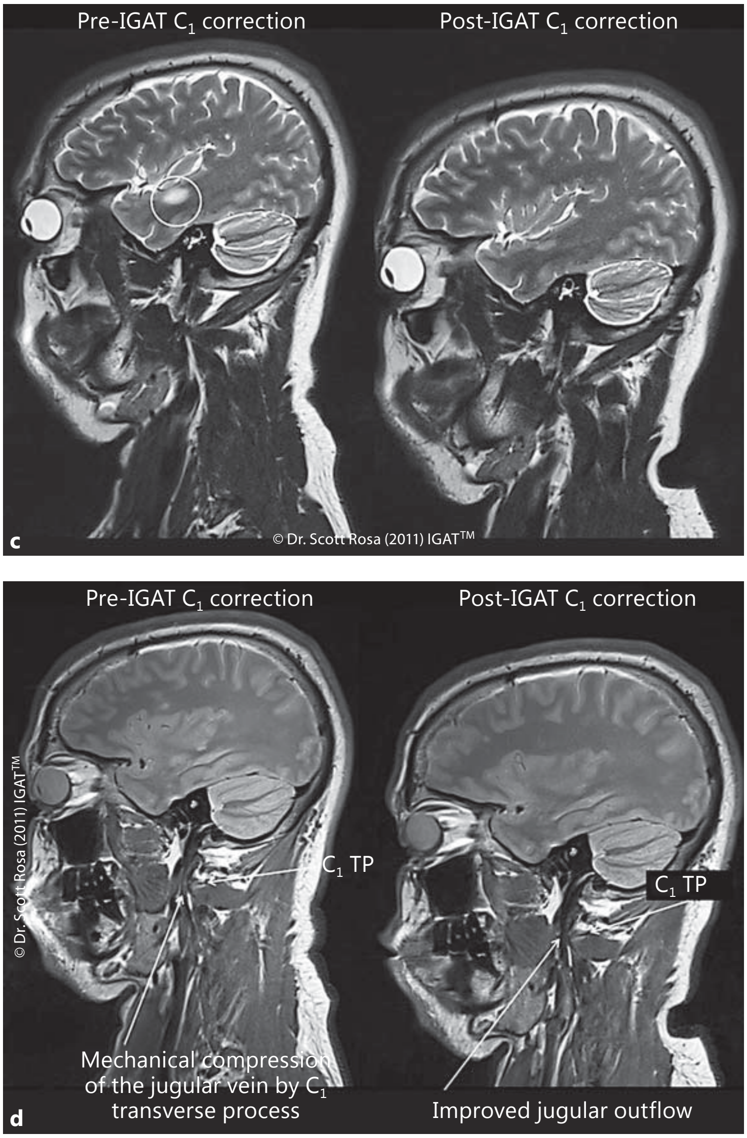

Fig. 15. Case 4: sclerotic lesions in a patient diagnosed with multiple sclerosis which have abated after IGAT C_1 correction (**c**). Mechanical compression of the jugular vein by the C_1 transverse process which improved after IGAT C_1 correction (**d**).

bleeding), but all too often the soft tissue structures inherently capable of manifesting in pain are overlooked [18]. The importance of understanding the nociceptive capabilities which these structures possess should not be underestimated.

Familiarity with all the soft tissue structures of the CCJ (ligaments/membranous structures and proper alignment) enables the assessor to identify plausible red flag signs that correlate with the patient's complaints. These red flag signs are illustrated in figures 1–9, and also include kyphotic angulation [19], paradoxical interspinous fanning [20] and listhesis on flexion/extension [21].

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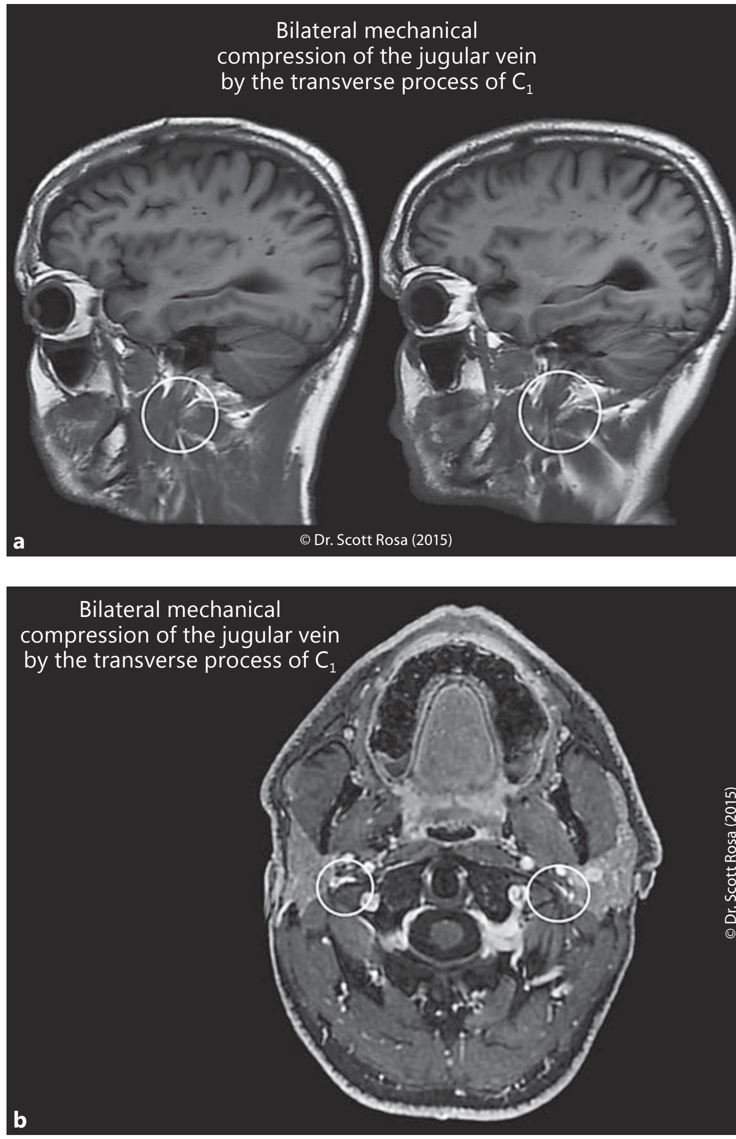

Fig. 16. Case 5: extravascular compression has been associated with chronic venous insufficiency and multiple sclerosis. The primary cause is mechanical compression by the C_1 transverse process [32]. **a**, **b** Bilateral mechanical compression of the jugular veins by the C_1 transverse process.

Pain Generators

Nociceptive structures at the CCJ have a fairly common pathway. Most of the pain receptors are mediated by the trigeminocervical nucleus, and are initiated by noxious stimulation of the endings of the nerves that synapse on this nucleus, by irritation of the nerves themselves or by disinhibition of the nucleus. Trigeminal nerve afferents will descend to the level of C_3 and perhaps as low as C_4 [22].

Important structures innervated by C_1–C_3 include the vertebral arteries, the carotid arteries, the alar ligaments (fig. 10a, b) and the transverse ligaments (fig. 11a, b).

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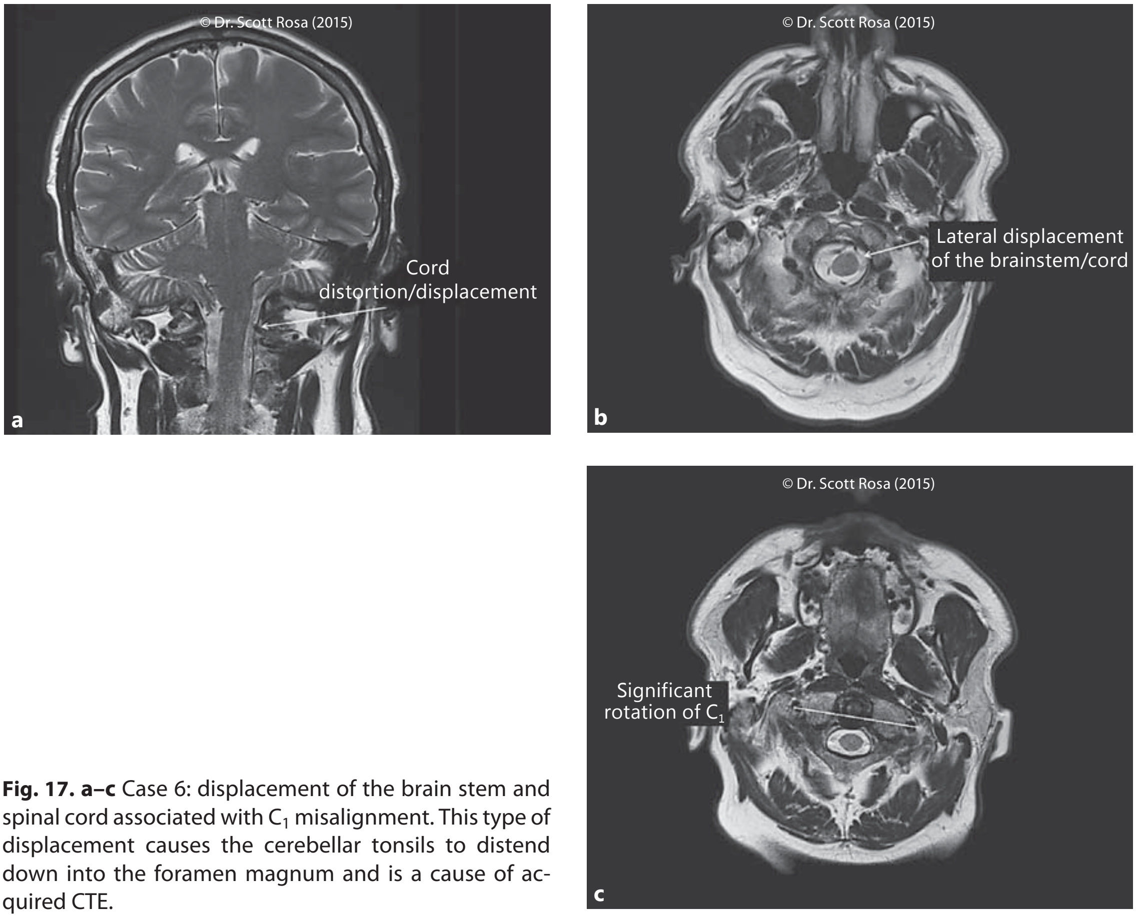

Fig. 17. a–c Case 6: displacement of the brain stem and spinal cord associated with C_1 misalignment. This type of displacement causes the cerebellar tonsils to distend down into the foramen magnum and is a cause of acquired CTE.

Spinal Pain

The atlanto-occipital, atlantoaxial and C_2/C_3 joint capsules may generate pain into the upper neck and also have the potential to cause headaches. Each joint has a unique pain referral pattern [23]. The myodural bridge attaching suboccipital muscles to the dura mater can be a cause of occipital headaches [24].

The dura is attached to the transverse ligament of the atlas and to the posterior longitudinal ligament, dorsally to the periosteum of the occipital squama and arches of the atlas and axis, and laterally to the atlanto-occipital and atlantoaxial articulations. The dura has nociceptive capabilities as well, and as such needs to be viewed on sagittal imaging data sets, especially as it courses posteriorly down and through the foramen magnum between C_0 and C_1. Dural infoldings on sagittal MRI at the CCJ have been identified in patients with flexion extension injuries and can be a source of headache and suboccipital pain.

Image-Guided Atlas Treatment

Image-Guided Atlas Treatment™ (IGAT™) utilizes dynamic upright MRI to permit the proper visualization of the CCJ misalignments. This en-

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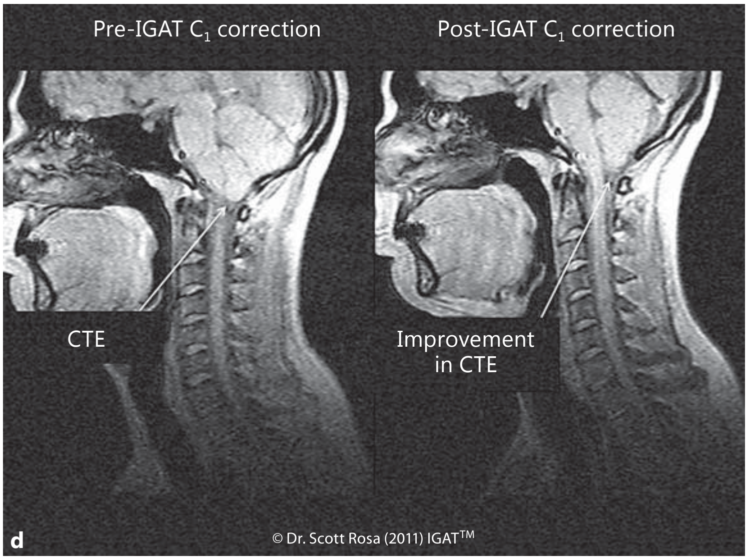

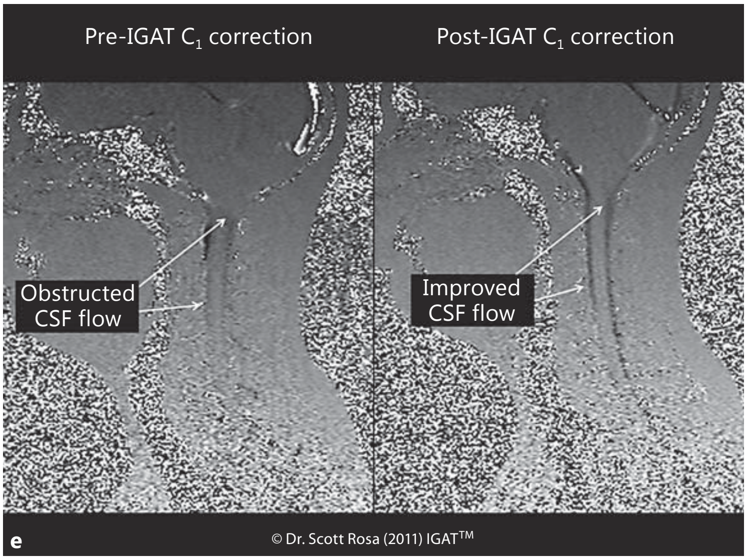

Fig. 17. d, **e** Case 6: CTE which is improved with IGAT C_1 correction. **e** Obstructed CSF flow which is improved with IGAT C_1 correction.

ables the ability to analyze and correct the misalignments in a nonmanipulative, gentle and safe manner. The correction is performed utilizing the 'atlas orthogonal percussion instrument'. This patented method of correction has been researched, developed and administered by Dr. Scott Rosa. It is based on a 'trauma injury protocol', or TIP, which uses advanced imaging methods for patients with mild, moderate and severe neck injury caused by various assaults to the spine. These include, but are not limited to, motor vehicle accidents, falls, sports and work-related industrial accidents. Injuries to the neck, or cervical region, are very important since there is a potential risk of insult to the spinal cord. Any neck injury can have devastating as well as life-threatening consequences [25].

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The cervical spine is a complex mechanism from a mechanical and structural point of view, acting as a tension-compression structure. Therefore, cervical spinal injuries are a potential threat to the spinal cord and must be treated with respect and caution. Cervical spinal injuries are often caused by impacts going beyond the physical threshold of ligaments and bone in the area, resulting in direct physical trauma. Spinal injuries, whether due to trauma, degenerative changes or disease, are extremely important to diagnose, treat and rehabilitate in the most precise and conservative manner possible. A series of three IGAT cases are presented in figures 12–17.

Discussion

Dynamic upright MRI is an invaluable tool to assess the CCJ. The imaging of this area must include sagittal, coronal and axial views for proper assessment. The treating physician should be cognizant of injury/disruptions in the stabilizing soft tissues of the area. It is no longer acceptable to just have a sagittal imaging sequence and then do axial images from the C_2 disc down. With the literature being replete with information discussing the myriad of symptom manifestation from the CCJ, it would be a disservice to the patient to not fully image the CCJ in a 3D plane, especially considering the 'craniocervical syndrome' and its entire reported plausible symptom picture. CSF flow studies are of great clinical importance and can ensure proper diagnosis, rather than a finding of 'normal MRI' due to a lack of obvious disc pathology. Assessment of spinal stability will determine the choice of treatment in each specific type of soft tissue injury. Correct evaluation of joint biomechanics, especially those in the cervical spine area, is paramount [26]. However, static diagnostic testing alone often does not have sufficient sensitivity to identify these nondisc-related soft-tissue injuries [27, 28]. Misdiagnosis may lead to complications in an otherwise treatable patient. With the utilization of dynamic imaging, specifically dynamic upright MRI, cervical spine hypermobility, hypomobility and instability, most often due to various traumas, can be identified and treated accordingly. Unstable spinal injuries may directly damage the spinal cord itself if not evaluated and diagnosed properly in the clinical setting. Flexion and extension views on upright MRI also permit improved visualization of alignment, motion and disc pathology over traditional static MRI images [29–31].

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