External syrinx-introduction of a new term

Syringomyelia has been evaluated, researched, and treated for over a century. A number of studies from worldwide have helped to clarify the subject. Despite the publication of several hundred articles on the subject that are available on PubMed/MEDLINE, controversy regarding its pathogenesis and rationale of its treatment continues. Syringomyelia is a chronic or longstanding and unrelentingly progressive collection of cerebrospinal fluid (CSF) within the intramedullary spinal compartment.[1] Chiari 1 malformation and syringomyelia are common associations. Basilar invagination and bone malformations at the craniovertebral junction are frequently associated with Chiari 1/syringomyelia complex.[1,2] We have speculated that both Chiari 1 malformation and syringomyelia are protective natural responses in the face of longstanding atlantoaxial instability.[3-5] Although Chiari 1 malformation is like a nature's airbag, placed in position to provide a cushion of protection to the vital part of the cervicomedullary cord against the abnormal movements of the odontoid process and compression between the bones, syringomyelia is a protective self-neural destruction that works in the overall favor of the neural structure and human survival.[3,4] Atlantoaxial fixation alone can result in reversal of both Chiari 1 malformation and syringomyelia.[5] This concept has the potential of dramatically altering the understanding of the pathogenesis of the complex and instituting the correct form of treatment.

Atlantoaxial joint is the most mobile joint of the body. Circumferential movements at the joint are facilitated by its flat and round articular surface structure. While the special bone and soft tissue architecture allow a range of movements, it also subjects it to the possibility of abnormal movements and instability. Our analysis of the subject reveals that atlantoaxial instability is an underdiagnosed and undertreated clinical entity. It seems likely that the atlantoaxial joint is the most unstable joint of the body. Atlantoaxial instability is traditionally evaluated by alteration of atlantodental interval. However, the understanding of vertical, lateral, central, or axial and rotatory atlantoaxial instability has added a new dimension to the subject.[6-12]

The large circumferential dimension of the ring of the atlas, larger than foramen magnum, is especially designed to permit free movement to the odontoid process and avoid neural compression. We labeled the "foramen" of ring of atlas as *foramen maxima*. The very fact that Chiari 1 malformation extends up to and seldom beyond the atlas suggests that tonsillar herniation is designed to protect the cervicomedullary cord from compression by the odontoid process against the bony posterior arch of atlas. The frequent presence of bifid posterior arch of atlas is a protective and natural "decompressive laminectomy."[13] We speculated that bifid arch of atlas provides an open-close door kind of phenomenon wherein the posterior arch opens upon flexion of the head to provide space to the odontoid process intrusion and closes on neck extension when the odontoid process is anteriorly positioned. The bone abnormality of the arch of atlas, lateral disposition of the facets of atlas in relationship with the facets of axis and occipital condyle, reduction of the height of the occipital bone and atlas and axis complex all seem to be protective natural designs instituted to protect the neural structures against potential damage by the abnormal movements of the odontoid process.[13] "Short neck," "short head" in the form of short clivus and posterior fossa bone compartment, torticollis, bone fusions, platybasia, and several other musculoskeletal malformations seem to be an effect of nature's attempt to reduce the length of the neural canal to permit a stretch free traverse of the brainstem and spinal cord.[14]

We have identified that there are several alterations in the spine and the posterior cranial fossa. The spinal canal increases in its circumferential dimensions and the spinal cord reduce in its girth. Similarly, the clivus becomes short

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resulting in the short head. Like the neck that becomes short and spinal canal that becomes larger, the posterior cranial fossa becomes shorter in its vertical height but longer in its transverse and anteroposterior dimensions. In addition, the neural structures in the posterior fossa that include cerebellum and brainstem become thin and atrophic. The overall effect increases the space available in the spinal canal and posterior cranial fossa that is occupied by CSF. In the spinal canal, the increased collection of CSF may be in the extramedullary space (external syrinx), inside the spinal cord (syringomyelia) or both inside and outside the spinal cord.[15] An excessive amount of CSF is also present in the posterior cranial fossa, inside the brainstem (syringobulbia), anterior and around the brainstem (external syringobulbia) and around the cerebellum.[15] The cerebellum, more in its superior vermis and superior cerebellum, becomes atrophic allowing an increased collection of CSF.[16] Superior vermis is atrophied, but the herniated part of the tonsil is never atrophied, probably because a firm cerebellar tissue is necessary to provide a compact cushion.

Atrophy of the neural structures at the point of compression by the odontoid process and in the spinal cord and brainstem seem to be a natural response to reduce the girth of neural structures that allows it to accommodate the intruding odontoid process and make the neural tissues resilient and stronger. Essentially, we speculated that the bony spinal canal and posterior fossa decrease in vertical height and neural structures (spinal cord, brainstem, and cerebellum) become thinner and longer in length, allowing the neural structures to course over the odontoid process in a relatively stretch free traverse. The direction of the angulation of the odontoid process influences the presence of CSF within or outside the neural tissues. We observed that more severe is the angulation of the odontoid process and more severe is the compression of the neural structures more is the incidence of the presence of external syrinx. Essentially, it seems that external syrinx and excessive collection of CSF in the posterior cranial fossa allows the neural structures to float away from the compressing odontoid process. Reduction of girth of the neural structures seems to be a result of escape of CSF in the central spinal canal and fluid inside and outside the neural fibers in the spinal cord. In this respect, it seems that the pathogenesis and function of CSF, whether it is inside the cord in the form of syringomyelia (or syringobulbia) or whether it is outside the spinal cord in the form of the external syrinx (or external syringobulbia) is essentially the same.[15] Syringomyelia is more often seen when the odontoid process is either in its normal position or is vertically herniated into the posterior cranial fossa. External syringomyelia is more often when the odontoid process is posteriorly angulated or horizontal in its lay. The marked resilience of the neural structures and preservation of the neural function despite the severe reduction of neural dimensions are unique protective ways of nature. Our observations suggest that atlantoaxial dislocation is the only craniovertebral anomaly. All other bony and soft tissue alterations are secondary to atlantoaxial instability and are protective in their function.

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