CONSENSUS STATEMENT

1. Ventral brainstem compression, medullary kinking and deformation of the upper spinal cord and/or brainstem over the odontoid process are potentially deleterious to the brainstem and upper spinal cord.

2. Deformation of the brainstem may manifest clinically as the Cervical Medullary syndrome.

3. The clinical findings of Cervical Medullary Syndrome may include, but are not limited to, the following:

i) headaches, suboccipital pain and neck pain,

ii) Bulbar and related symptoms: altered vision, diplopia, nystagmus, decreased hearing, tinnitus, imbalance, vertigo, dizziness, choking, dysarthria, dysphagia dysautonomia, postural orthostatic tachycardia, pre-syncopal or syncopal episodes disordered sleep architecture, sleep apnea,

iii) Symptoms of myelopathy: weakness, clumsiness, spasticity, altered sensation, paresthesias, dysesthesia, change in gait, constipation, urinary urgency and frequency

4. In assessing the potential for cranio-cervical instability, it is reasonable to measure the angle between the clivus and the spine. This angle has been termed the clivus canal angle, the clivus vertebral angle, the clivus spinal angle, the clivus cervical angle and the clivus-axial angle.

In keeping with the greater part of the literature, we recommend the uniform adoption of the term clivo-axial angle .This angle may be abbreviated CXA.

5. The clivo-axial angle is the angle between the clivus line and the posterior axial line. The clivus line is drawn along the lower third of the clivus -from the spheno-occipital synchondrosis to the basion, or in the case of basilar invagination, the superior most aspect of the odontoid .

When assessing the CXA with sagittal CT scan or X-ray, the posterior axial line may be drawn along the posterior edge of the odontoid .

When assessing the CXA with MRI, the posterior axial line should be drawn from the posterior edge of the tectorial membrane to the inferior posterior edge of the posterior ligament of the C2 vertebra.

The CT and MRI measurements may differ in the same patient: the CXA determined by CT reflects the more traditional means of measurement ; the CXA determined by MRI will necessarily include thickening of the ligament due to pannus.

6. The literature suggests that a clivo-axial angle of 135 degrees or less is potentially pathological. That is a CXA of135 degree, may in some circumstances, result in harmful deformative stress upon the brainstem and upper spinal cord and, therefore, warrants consideration for further evaluation and possible treatment.

7.The CXA can be measured on sagittal CT or MRI, with the patient assuming moderate flexion of the cranio-cervical junction. If a flexion view is not available, a neutral position will suffice in most circumstances. An upright dynamic MRI may be desirable in some circumstances – but, such is often not available.

8. In assessing the potential for cranio-cervical instability, it is reasonable and appropriate to measure the BpC2 line, also known as the Grabb-Oakes measurement or line, or the Grabb Mapstone Oakes Measurement, as one method to approximate the potential presence and magnitude of ventral brainstem compression. We use the term Grabb-Oakes measurement herein. The Grabb-Oakes measurement is the distance in millimeters from the dura to the line drawn from the basion to the posterior inferior edge of the C2 vertebra. A Grabb Oakes measurement of 9mm represents the diagnostic threshold for ventral brainstem deformity. Some clinicians may choose 8 mm as the diagnostic threshold at which there may is potential ventral brainstem deformity.

9. The Harris measurement, also known as the Basion Axial Interval (BAI = distance from tip of basion to posterior axial line), when drawn horizontally, should be less than 12 mm. The basion to dens interval (BDI = distance from basion to tip of odontoid) drawn vertically, should be less than 12 mm. The posterior axial line should be drawn along the posterior ligamentous surface of the C2 vertebra. In keeping with the literature, a Harris measurement exceeding 12 mm is considered potentially pathological, and reflects cranio-cervical instability.

10. In the presence of known ligamentous instability, such as a hereditary hypermobility connective tissue disorder, the BAI (the Harris measurement) may be measured with the cervical spine in the flexion and extension positions. This will assess and quantify translation of the basion with respect to the dens (odontoid process). In keeping with the literature, any translation noted on dynamic imaging that exceeds 2mm (the delta BAI > 2 mm), will be considered abnormal and potentially pathological.

11. Cranio-cervical hypermobility is common, and defined by the presence of hyper-extensibility of the connective tissue, and in particular, hyper extensibility of the joints. While hypermobile joints occur frequently in healthy children, such can also be severely disabling in others. Ehlers Danlos syndrome, cleidocranial dysostosis, Down syndrome, Marfan syndrome, Morquio syndrome and several other less well known connective tissue disorders are associated with ligamentous laxity. A pathological Lax Ligament Syndrome may result in cranio-vertebral instability, kyphosis of the clivo-axial angle and ventral brainstem compression.

The growing body of knowledge regarding the prevalence of hypermobility connective tissue disorders should lead to more widespread recognition of the impact of ligamentous laxity on the health of sufferers of hypermobility syndromes.