Diagnosis & Management of Syndromes of the Craniocervical Junction and Roundtable Discussion

2019 London Consensus Conference

13 September 2019

The Royal Society of Medicine
1 Wimpole Street
Naim Dangoor Auditorium
London, England

Summary of Meetings & Consensus Points
Presented by: Bobby Jones Chiari & Syringomyelia Foundation and The Ehlers-Danlos Society

Summary Consensus Document

A consensus conference was sponsored jointly by the Bobby Jones Chiari Syringomyelia Foundation, the Ehlers-Danlos Society, the Office of the Medical Commissioner of the National Health Service and the Lady Trish Malloch-Brown.

Lectures and arguments were put forward addressing the presentation of craniocervical instability, basilar invagination and co-morbid conditions, such as dysautonomia, postural orthostatic intolerance and migraine which occur in the population of patients with hereditary connective tissue disorders.

The lectures were followed by a robust “round table” discussion, enjoined by representatives of the neurosurgical and neurological, neuro-radiological, genetic, rheumatology and physiatry communities, to establish a consensus regarding the diagnosis and treatment of craniocervical disorders in this population.

The following discussion points were unanimously agreed upon:

1. That hereditary connective tissue disorders— including, but not limited to Marfan syndrome, the Ehlers-Danlos syndromes, Down Syndrome, osteogenesis imperfecta— are characterized by ligamentous incompetence;

2. That in a small subpopulation of subjects with an hereditary connective disorder, ligamentous incompetence may result in radiological evidence of craniocervical or spinal instability, basilar invagination or ventral brainstem compression;

3. That dynamic imaging is necessary to demonstrate ligamentous incompetence at the craniocervical junction; dynamic imaging may include flexion extension MRI, flexion extension or rotational CT, or in some cases fluoroscopic imaging;

4. That a diagnosis of radiological instability, basilar invagination or ventral brainstem compression should be characterized by radiological metrics which have been established in the literature;

5. That Subjects with hereditary connective tissue disorders who receive a radiological diagnosis of craniocervical instability, basilar invagination or ventral brainstem compression should undergo a neurosurgical evaluation for the presence of clinical findings of instability;

6. That Subjects with appropriate clinical findings and congruent radiological findings of craniocervical instability, basilar invagination or ventral brainstem compression, who meet standard surgical indications should be considered for craniocervical, or atlanto-axial, reduction, stabilization and fusion;

7. That members of this colloquium elect a panel to work with the National Health Service to commission the use of appropriate imaging techniques in Subjects suspected of harboring craniocervical instability, basilar invagination or ventral brainstem compression, to confirm or rule out the diagnosis.