Chiari malformation Type 1 and atlantoaxial instability: a letter from the Pediatric Craniocervical Society

TO THE EDITOR: In his article, Dr. Atul Goel examines the underlying cause and treatment of Chiari malformation Type I (CM-I) in 65 patients at his institution in Mumbai, India (Goel A: Is atlantoaxial instability the cause of Chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. J Neurosurg Spine 22:116–127, February 2015). All were treated with a surgical technique to fuse the atlas and axis without foramen magnum decompression or intradural exploration. Sixty-three of the patients had favorable outcomes, 1 failed to improve after surgery, and 1 died from a vertebral artery injury. The atlantoaxial fusions were performed on the basis of Dr. Goel’s novel classification scheme, with or without any radiographic evidence of instability of the joint. At the conclusion of the paper, Dr. Goel suggests that all patients with CM-I be treated with this technique.

After carefully reviewing the paper, we believe that adopting this approach for all patients with CM-I is wrong and potentially dangerous.

Dr. Goel’s patient population was complex, with a high representation of craniovertebral junction anomalies and large syringes, atypical of the general patient population with CM-I. Basilar invagination was identified in 46 of 65 patients and syringomyelia in 55 of 65 patients. Only 11 of 65 patients were “independent and normally functioning,” with 12 of 65 “unable to walk and dependent for all activities.” This particular patient group is more advanced in their symptoms and more complex than those who are typically seen at other centers. Historically, these patients are difficult to successfully treat and do not represent the majority of patients with CM-I. Authors from multiple centers worldwide have reported excellent outcomes for patients with CM-I who are treated using foramen magnum decompression and dural opening without atlantoaxial fusion. Therefore, the suggestion that we abandon these traditional operative techniques is premature.

Defining instability in these complex patients can be challenging. It is well known and accepted that occipitocervical fusion is necessary in a small subset of complex patients with CM-I, but applying these techniques to all patients with CM-I, especially those without evidence of spinal instability, is illogical. Dr. Goel’s novel classification scheme of “atlantoaxial dislocation” has not been confirmed, standardized, or validated. One of Dr. Goel’s specific assertions is that one can restrict fusion to the atlantoaxial level and have the same effect as an occipitocervical fusion. This argument is undermined by the fact that, as pointed out in Dr. Jea’s editorial, at least 33 of 65 patients had occipitalization of the atlas, and thus essentially turning Goel’s atlantoaxial fusions into the biomechanical equivalent of an occipitocervical fusion. Five of the 6 illustrated cases in the paper demonstrate this point.

To convince others to adopt his novel approach, Dr. Goel will need to confirm his findings with rigorous postoperative assessment. In his paper, only 11 of 65 patients had postoperative MRI, and 5 of 11 apparently showed no syrinx change. Although 1 death was reported, an additional patient had “clear evidence of a posterior circulation infarct related to an intraoperative vertebral artery injury.” It is not clear whether this patient was in the group that improved postoperatively.

Many questions remain unanswered: 1) What specific criteria were used to pick the patients for atlantoaxial fusion? 2) Why did patients undergo fusion without traditional evidence of atlantoaxial instability? 3) What clinical and radiographic criteria were used to define success? 4) Applying Dr. Goel’s methodology and conclusions, how does he explain the excellent reported outcomes in the thousands of pediatric and adult patients with CM-I who underwent foramen magnum decompression without atlantoaxial fusion?

It is clear that there is a small subgroup of patients with CM-I and significant craniovertebral junction anomalies that may benefit from an occipitocervical fusion when instability is present; however, until Dr. Goel demonstrates that his novel classification system and clinical management are appropriate and applicable to the majority of patients with CM-I, universal application of his strategy of atlantoaxial fusion without foramen magnum decompression to the general CM-I population is unwarrented and may be potentially harmful.

Douglas L. Brockmeyer, MD
University of Utah, Primary Children’s Hospital, Salt Lake City, UT

W. Jerry Oakes, MD
Curtis Rozzelle, MD
James Johnston, MD
Brandon G. Rocque, MD, MS
University of Alabama, Birmingham, AL

Richard C. E. Anderson, MD
Neil Feldstein, MD
Columbia University Medical Center, New York, NY

Jonathan Martin, MD
Connecticut Children’s Hospital, Hartford, CT

Gerald F. Tuite, MD
Luis Rodriguez, MD
University of South Florida/All Children’s Hospital, St. Petersburg, FL
assimilation of the atlas, secondary spondylotic changes, and several other musculoskeletal events associated with basilar invagination may not be congenital anomalies, but rather Nature’s protective responses in the face of atlantoaxial instability. Atlantoaxial fixation has resulted in the reversal of several of the above-mentioned physical abnormalities in the immediate postoperative period.26,30 We also mentioned that there is potential for the reversal of all musculoskeletal events including bone fusions.30 We now observe that both CM and syringomyelia in the presence or absence of basilar invagination are a continuum of the same phenomenon and are natural events that are protective. While CM simulates an air bag, syringomyelia is a natural self-neural destructive process that works in the greater interest of the body. Both CM and syringomyelia should be respected as protective, not as pathological events, and should be treated as such.11,13

Our experience in dealing with and manually handling unstable atlantoaxial and subaxial spinal facets is more than 25 years long.1–7,10,13–20,22–29,31–36 We have treated over 1300 cases by atlantoaxial fixation.22,26 Because of this unique opportunity, we are able to identify the presence or absence of instability rather easily by assessing the local stability situation during an operation by manual bone handling. Atlantoaxial instability has traditionally been diagnosed by assessing the atlantoaxial interval and by observing its alteration on dynamic imaging. Although the concept of “facetal” instability, as proposed by us, is neither established nor validated, it appears to be a phenomenon that opens up space for a fresh understanding of the subject.8,9 The fact that instability can be assessed by facetal alignments can have a great impact on understanding the dynamics of the region and in conceptualizing the treatment options. In cases with basilar invagination, we had earlier identified anterior dislocation of the facet of the atlas over the facet of the axis and labeled it atlantoaxial listhesis, a situation similar to lumbosacral listhesis.10,17,19,36 In this situation the atlantoaxial interval increases, and there is direct indentation of the neural structures by the odontoid process. The clinical presentation in such cases is rather acute, and chronic musculoskeletal and neural alterations are not the hallmark of such cases. We labeled this type of facetal instability as Type I dislocation. We further identified instability of the atlantoaxial region as manifested by posterior dislocation of the facet of the atlas over the facet of the axis and labeled it atlantoaxial listhesis, a situation similar to lumbosacral listhesis.10,17,19,36 In this situation the atlantoaxial interval increases, and there is direct indentation of the neural structures by the odontoid process. The clinical presentation in such cases is rather acute, and chronic musculoskeletal and neural alterations are not the hallmark of such cases. We labeled this type of facetal instability as Type II dislocation. We further identified instability of the atlantoaxial region as manifested by posterior dislocation of the facet of the atlas over the facet of the axis (Type II). We also identified Type III atlantoaxial facet instability, wherein the facets are in alignment but the instability is identified by corroborative clinical and radiological evidence and observed only by direct intraoperative handling of the facets. We called such instability “central” or “axial” atlantoaxial instability. The atlantoaxial interval is unaffected in both Types II and III, and direct compression of the neural structures by the odontoid process is not evident. Moreover, instability in these cases is rather subtle, although it is definite and pathological. Essentially, Type II and Type III atlantoaxial facet dislocations are associated with longstanding or chronic events such as Group B basilar invagination and/or CM with or without the association of syringomyelia.

In the year 2004, we demonstrated that the atlantoaxial joint in select cases of basilar invagination (Group

Response

It is clear from this letter (and from previous correspondences) that my article has been received with great interest. While I am convinced about the various theories I proposed and my continuing clinical experience reaffirms my belief, the letter provides me with an additional opportunity to present my viewpoint to the readers and to the various experienced and respected neurosurgeons listed in the letter. At variance with the decades-old concept that CM is a pathological entity and an event related to embryonic dysgenesis, my article suggests that CM could be Nature’s protective phenomenon and that tansillar herniation simulates an air bag.11 The air bag is positioned to cushion the critical craniovertebral cord against impending pinching between bones in the event of local instability. The instability seems to be recognized by Nature, protective acts are initiated early in life, and the clinical manifestations are delayed. We had earlier hypothesized that a short neck, torticollis, platybasia, Klippel-Feil anomalies,