Editorial

Chiari malformation Type I and scoliosis: the complexity of curves

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“One finds in the study of the origin of most pathological conditions that they are at first confused with other similar affections, gradually emerging and being identified. In scoliosis this is particularly the case.” So states Lovett⁴ in his 1913 historical treatise on scoliosis, which may be read as a blueprint for the eventual recognition and identification of Chiari malformation Type I (CM-I)–related spinal curvature. Indeed, since that time, much has been learned regarding CM-I-related scoliosis, yet most surgeons would admit that much remains unknown. Patients presenting with CM-I and scoliosis, with or without syringomyelia, can create some of the most challenging clinical scenarios in pediatric neurosurgery. Therefore, studies that can provide further understanding of this association are most welcome.

The association between CM-I, scoliosis, and syringomyelia in children was first described in 1988 by Dausser et al. To this day, a definitive explanation of the proximate cause of CM-I-related scoliosis remains elusive. That all Chiari-related curves are pathological compared with idiopathic curves suggests that a unique mechanism is responsible. Chiari malformation Type I–related scoliosis is characterized by lack of coronal balance, a tendency toward single-apex curves, and a higher incidence of left thoracic apices. Some authors believe that dorsal tonsillar compression on the cervicomedullary junction initiates and propagates the scoliosis, while others believe that syringomyelia is the driving force. It is also thought that asymmetric cross-sectional syrinx formation may lead to asymmetric anterior horn cell dysfunction, paraspinous muscle imbalance, and scoliosis. The latter explanation is hard to understand, however, when CM-I-related scoliosis occurs in the absence of syringomyelia, as has been reported. Small posterior fossa volumes are also found at a higher frequency in patients with CM-I who are symptomatic and have syringomyelia or scoliosis. With such a wide variety of possible explanations, it is clear that the events of CM-I-related scoliosis initiation and propagation are not yet fully understood.

In their article in the current issue of Journal of Neurosurgery: Pediatrics, Krieger et al.⁵ present a large retrospective case series documenting a single institution’s experience in managing CM-I-related scoliosis. They should be congratulated on their excellent results; however, based on previous work, these results are not surprising. The authors confirm that patients who presented with Cobb angles less than 20° or at an earlier age fared better than those presenting with curves over 20° or in adolescence. These findings are similar to those reported by Eule et al., Brockmeyer et al., and Attenello et al. Krieger et al. also report that there was almost complete resolution of syringomyelia in a high percentage of their cases. These results help to solidify the notion that early recognition of atypical scoliosis is important and leads to identification of a fairly high percentage (approximately 10%) of patients in whom the scoliosis is caused by a CM-I. Additionally, timely treatment of the hindbrain compression leads to favorable outcomes of CM-I-related scoliosis and syringomyelia in a majority of cases.

Several relevant questions are left unanswered by this article. To begin with, Krieger et al. describe their own technique of suboccipital decompression for these cases, which consists of a suboccipital craniectomy, C-1 laminectomy, dural opening, and arachnoid dissection without dural closure. Can this technique be applied across other patient populations as well? Does the dura have to be opened? Does it have to be closed? Should the cerebellar tonsils be dissected free and reduced? The authors’ results, along with those from other reports, suggest that patients do best when the dura is opened and the arachnoid is lysed. Furthermore, tonsillar shrinkage was not mentioned, which leaves the reader with the impression the authors believe that it is not a necessary part of the procedure. For many surgeons, including myself, tonsillar reduction is a standard part of the procedure when patients present with scoliosis. Whether the dura needs to be closed or not is another issue. Most surgeons go out of their way to perform a watertight dural closure, often with duraplasty, in the posterior fossa. The authors’ experience suggests that this may not be necessary, although they had a 9% rate of CSF wound leakage and a 16% rate of aseptic meningitis. Our own rates of 0% CSF leakage and aseptic meningitis over our last 22 CM-I decompressions for scoliosis seem to suggest that dural closure may not be a bad idea.

One of the more vexing problems in this patient population is the management of a residual syrinx after an adequate CM-I decompression for scoliosis. The authors report a 13% rate of residual syringomyelia in their study, which is in keeping with our own results. They also report that by either placing a syringoperitoneal shunt or
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performing a reexploration of the CM-I decompression, all syringes eventually came under “control.” Since syringomyelia might be the driving force behind scoliosis in the majority of cases, it stands to reason that resolution of the syrinx should be a high priority. Also (and this has never been studied), we do not know the long-term effects of syringomyelia on the spinal cord and its role in producing neurological deficits, pain syndromes, and chronic debilitation in adult patients. So is aggressive management of syringomyelia justified in the pediatric population? If one assumes that the syrinx is causing the scoliosis, the answer appears to be yes. Unfortunately, the authors do not separately report the outcome of scoliosis in the 10 patients with residual syringomyelia, presumably because they came under “control.” This would be valuable information from such a large series of patients.

One notable exclusion by the authors is any kind of description of ventral brainstem compression, CM Type 1.5, or congenital vertebral anomalies in their patient population. In our own series of patients with CMs, these findings are relatively common and have garnered the moniker “complex Chiari.” While the relative importance and clinical significance of these findings can be debated, in our experience they have become very important for our clinical decision-making process. For example, in the last 22 patients who presented with scoliosis and syringomyelia, 7 had a CM Type 1.5 and 6 had significant ventral brainstem compression due to retroflexed odontoid (defined as a pBC line > 10 mm [with pBC2 as the line drawn from the tip of the odontoid to the ventral aspect of the dural]). These findings sometimes lead to a complex scenario in which the surgeon must decide whether ventral compression is contributing to the hindbrain compression and whether reducing it (via a posterior or anterior procedure, or both) is desirable or necessary. One patient in our series with scoliosis and syringomyelia had a CM Type 1.5 and ventral brainstem compression (pBC of 10 mm). After standard decompression of the CM, tonsillar shrinking, and duraplasty, the malformation recurred and the syrinx failed to resolve. After reexploration of the CM and tonsillar shrinking, combined with odontoid resection during posterior occipitocervical fusion, the syrinx resolved and scoliosis improved. This case is mentioned not to sanction a particular way of management but to illustrate how complex some of these patients can be and how little we know about the “correct” way to manage them.

The case series presented by Krieger et al. seems to suggest that all patients with a CM-I, scoliosis, and syringomyelia are similar and can be managed in a standardized fashion. In reality, this is a very diverse patient population and managing their complex problems requires a large skill set and an open mind. It seems that the topic of CM, scoliosis, and syringomyelia would be well suited for a multicenter study comparing operative techniques and outcomes. I would suggest that further energy regarding this topic should be channeled in that direction because, with our less-than-perfect track record in managing CM-related scoliosis and syringomyelia, we, as Lovett6 reminds us, still have a long way to go in understanding this fascinating condition.

Response

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We thank Dr. Brockmeyer for his comments regarding our paper.

We agree with Dr. Brockmeyer that, although there is a definite association between scoliosis and syringes, the cause and effect are less than clear. An asymmetric cross-sectional syrinx, that is, hydrosyringomyelia, affecting the neuronal input to the paraspinal muscles unequally seems intuitively to be the more likely pathophysiological explanation in many cases. Dr. Brockmeyer notes that only 10% of scoliosis cases are associated with a CM-I. It may be that those pediatric patients with a CM-I and no hydrosyringomyelia with scoliosis may have scoliosis that is not related to the CM-I.

Dr. Brockmeyer questions 3 elements of our described standardized operative procedure: dural closure, dural opening, and tonsillar resection. Dural closure adds 2 elements to the operative procedure—time and a foreign body, with the latter being more relevant. Although our experience may be skewed, we have noted considerable local reaction to dural substitutes at the time of a reoperation, whereas little such reaction is observed when no substitute has been placed. A “watertight” dural closure is a relative term. If CSF pressure is elevated, the closure will leak and if not, the watertight closure is largely irrelevant. We have typically placed subcutaneous dissolvable sutures to produce a better cosmetic closure. The CSF leakage, 9% of the series, was addressed by placing additional cutaneous sutures at the bedside in all patients.

References