Tethered cord

To the Editor: I read with great interest the article by O’Neill et al. (O’Neill BR, Yu AK, Tyler-Kabara EC: Prevalence of tethered spinal cord in infants with VACTERL. Clinical article. J Neurosurg Pediatr 6:177–182, August 2010). We are very pleased to see that the authors have confirmed our previously published findings and encourage healthcare providers to screen patients with the VATER association.1

We disagree with the authors, however, regarding their conclusion that MR imaging should be the procedure of choice in infancy and early childhood, even in this disease process and potential occult spinal dysraphia. Despite other publications to the contrary,2 we have previously published the advantages of spinal ultrasonography over other forms of studies in infants.3 These advantages include, but are not limited to, obtaining real-time images that can be obtained by no other means; being able to actually visualize the pulsations of the conus and cauda equina in the subarachnoid space; being able to perform the studies without any sedation or anesthesia (because the infant can move during the study). The major advantage of ultrasonography in real time, aside from the anatomical imaging, is that lack of pulsations of the conus and cauda equina can be readily seen during the study and is a powerful indicator of cord tethering.

Overall, the impact on reducing healthcare cost and ability to minimize exposure to anesthetic or sedative agents are very important factors. Unfortunately, ultrasonography is highly operator dependent. It is valuable for neurological surgeons to see real-time images themselves, which can be readily done with video systems. Real-time spinal ultrasonography has become very important in other areas of neurosurgery, such as single-level rhizotomy for spasticity.

Perhaps one solution to increase the knowledge of spinal ultrasonography is for fellowship program directors in pediatric neurological surgery to emphasize the exposure of the fellows during their fellowship training.

In no way do we wish to detract from the important information provided by O’Neill et al. as to the significance of studying children with VACTERL spectrum for occult spinal dysraphism.

Although we employed a 10-mHz transducer in our initial report,4 the current has improved to the extent that these studies can now be performed with almost any currently available ultrasonography equipment.

Hector E. James, M.D.
University of Florida College of Medicine
Jacksonville, Florida

Disclosure

The author reports no conflict of interest.

References


Response: I greatly appreciate the thoughtful comments shared by Dr. James and acknowledge his role in identifying the correlation between VACTERL and spinal tethering. I also appreciate the opportunity to respond to his comments.

It was notable in reviewing the literature on spinal ultrasound how few reports (less than 300 patients studied) correlate ultrasound to spinal MR imaging and how different the conclusions of these reports are. The largest series that we identified included 50 patients.1 This report found a very good overall correlation, with 96% sensitivity when dichotomizing the results to “normal” or “abnormal.” The authors did find less than 50% sensitivity in identifying the 13 dermal sinuses in the study and had only 4 fatty fila, all of which were correctly diagnosed on ultrasound. In contrast to this positive result, the study by Hughes et al.2 that Dr. James referenced found full agreement between ultrasound and MR imaging findings in only 40% of their 15 patients. Ultrasound missed 4 of 4 dermal sinuses, 3 of 4 fatty fila, and 3 of 4 syringes. Their reviewed population was high risk, with 12 of the 15 MR imaging studies displaying abnormalities. In our cohort, 35 patients underwent both ultrasonography and MR imaging of the spine. Of the 14 with tethered spinal cord who had ultrasonographic examinations, 3 had findings reported as normal. Admittedly, this cohort is at very high risk for spinal cord tethering and those patients with negative ultrasonographic studies who underwent MR imaging are a selected group. Still, the rate of false-negative results is concerning.

Dr. James espouses the value of assessing conus and filum pulsations on spinal ultrasound. The above-mentioned studies fail to evaluate this technique independently, but one wonders whether this or other technical factors account for the discrepant results (Ben-Sira et al.3 specifically mention that assessment for pulsation is performed while Hughes does not address the subject). A more provocative question is whether the functional information of a non-pulsatile conus better reflects the pathophysiology.
and whether it predicts the development of clinical symptoms.

We fully agree that the reduced cost of ultrasonography as compared to MR imaging and the avoidance of exposure to anesthesia are important factors that favor the use of this technology; however, the published information about ultrasonography’s accuracy raises concern about its value as a screening tool. Further study directly comparing ultrasonography to MR imaging is warranted.

References


Endoscopy or microscopy?

To the Editor: In their article, Kalani et al. (Kalani MYS, Kalani MA, Kalb S, et al: Craniofacial approaches to large juvenile angiofibromas. Clinical article. J Neurosurg Pediatr 8:71–78, July 2011) presented their experience with craniofacial resection for juvenile nasopharyngeal angiofibromas (JNAs) with intracranial extension. Their preferred approach was an anterior transbasal approach with a transmaxillary approach. In a series of 22 patients with intracranial tumor (Radkowski Stage IIIIB: extensive intracranial extension ± cavernous sinus involvement), gross-total resection was achieved in 17 (77%) and the average duration of hospitalization was 8.2 days. Complications were seen in 41% of patients; CSF leak was the most common complication, occurring in 23% of patients. The rate of recurrence and/or progression of disease was 18%; 3 patients required additional surgery and 5 patients received postoperative radiation therapy. The authors concluded that craniofacial resection of large JNAs is a preferred route and has significant but acceptable morbidity. Furthermore, they state “although endoscopic resection can be performed for small lesions, open approaches remain the standard of care for large JNAs.” The authors are to be commended for providing a detailed and honest description of their experience. We disagree, however, with their selection of the optimal surgical approach and their overall treatment strategy.

Current staging systems for JNA categorize intracranial extension as the highest stage. The amount of intracranial extension is not the most important prognostic factor for surgery; rather, it is the remaining tumor vascularity following embolization. This is reflected in the staging system proposed by Snyderman et al.1 In the absence of residual vascularity, most JNAs are readily dissected from the dura mater without the need for extensive exposure. Even with residual vascularity following embolization, we and others have successfully employed endoscopic endonasal and transfacial approaches with excellent results for even the largest of tumors, regardless of intracranial extension.

In our opinion, the craniofacial approach espoused by the authors does not provide optimal exposure and results in excessive morbidity. In our experience, most intracranial JNAs involve the middle cranial fossa and access the cranial cavity through skull base foramina (orbital fissures, foramen rotundum, foramen ovale). In such cases, a lateral infratemporal skull base approach provides better access to the middle cranial fossa and petrous internal carotid artery than a subfrontal approach.3 Tumors that erode the skull base medial to the cavernous internal carotid artery are best approached using a midline approach. A midfacial degloving approach augmented with a maxillectomy provides both medial and lateral access for large tumors. A LeFort I osteotomy or palatal split has not been necessary. Additional potential morbidities of a craniofacial approach not addressed in the paper are loss of olfaction, diplopia from disruption of the trochlea, and asymmetry of the orbits from detachment of the medial canthal ligaments.

Although the risk of residual or recurrent tumor remains high for advanced tumors (7 of 14 UPMC [University of Pittsburgh School of Medicine] Stage IV/V tumors), the majority of patients with small foci of residual intracranial tumor can be observed without additional therapy.3 In our series, 2 of 5 patients with residual tumor had progressive growth and were treated with endoscopic surgery. None of our patients have required radiation therapy, and its use should be discouraged in this young population. When revision surgery is necessary, it can be accomplished using endoscopic techniques with minimal morbidity.

With endoscopic endonasal and transmaxillary approaches, large JNAs with intracranial extension can be safely removed with equivalent tumor control and minimal morbidity. Potential benefits include improved visualization of the skull base, the avoidance of morbidities associated with craniofacial resection, shorter hospitalization, and faster recovery. We respectfully posit that craniofacial resection and postoperative radiation therapy are no longer the standard of care. Rather, a combination of extracranial approaches tailored to the location, routes of extension of the tumor, and residual vascularity of the tumor are preferred. If an experienced skull base team is available, these goals can be accomplished using endoscopic or endoscopic-assisted techniques.

(http://thejns.org/doi/abs/10.3171/2011.8.PEDS11337)

Carl H. Snyderman, M.D., M.B.A.
Paul A. Gardner, M.D.
Juan C. Fernandez-Miranda, M.D.
University of Pittsburgh School of Medicine
Pittsburgh, Pennsylvania

Disclosure

Dr. Snyderman is a consultant for Phacon.

References