

Evidence Discussion (HD-5)

Chiari and Skull-Base Malformations

- A CT scan of the head is less sensitive than an MRI of the brain for evaluation of intracranial structures, including major structural abnormalities of the posterior fossa.^{13,14}
- Neuroimaging in the initial evaluation of Chiari malformation should include the spinal cord due to the common occurrence or increased frequency of associated conditions such as cervical syrinx and tethered cord.^{1,2,3,4,9,10}
- For initial evaluation, treatment planning, and follow up, MRI is the preferred modality for malformations of the brain and cervicocranial junction.^{7,13,14,15} MRI is ideal for evaluating soft tissues, neural structures, and ligaments.^{7,11}
- As congenital brainstem and cerebellar anomalies are associated with spinal anomalies, MRI of the complete spine is helpful for diagnosis, follow up and treatment planning.^{1,2,3,4,13}
- A phase-contrast CSF flow study at the craniocervical junction is a supportive study for evaluation of Chiari malformation.^{5,15}
- Evaluation of cervicocranial junction anomalies, including basilar invagination and platybasia, may require more than one modality for diagnosis and surgical planning.^{7,8,11} CT characterizes osseous anatomy and may be helpful for surgical planning.^{7,11} MRI is preferred for evaluation of the soft tissues, neural structures and ligaments for these conditions.^{7,11} As craniocervical junction anomalies may lead to compression of adjacent vascular structures, CT- or MR-Angiography of the head and neck are useful for surgical planning.¹¹

References

1. Strahle J, Muraszko KM, Kapurch J, et al. Chiari malformation Type I and syrinx in children undergoing magnetic resonance imaging. *J Neurosurg Pediatr.* 2011 Aug; 8 (2): 205-213
2. Strahle J, Muraszko KM, Kapurch J, et al. Natural history of Chiari malformation Type I following decision for conservative treatment. *J Neurosurg Pediatr.* 2011 Aug; 8 (2): 214-221
3. Strahle J, Muraszko KM, Garton HJL, et al. Syrinx location and size according to etiology: identification of Chiari-associated syrinx. *J Neurosurg Pediatr.* 2015 July; 16 (1): 21-9 Epub 2015 Apr 3
4. Strahle J, Smith BW, Martinez M, et al. The association between Chiari malformation Type I, spinal syrinx, and scoliosis. *J Neurosurg Pediatr.* 2015 Jun; 15 (6): 607-611
5. Victorio MC, Khoury CK. Headache and Chiari I Malformation in Children and Adolescents. *Seminars in Pediatric Neurology.* 2016;23(1):35-39
6. Radic JAE, Cochrane DD. Choosing Wisely Canada: Pediatric Neurosurgery Recommendations. *Paediatrics & Child Health.* 2018;23(6):383-387. doi:10.1093/pch/pxy012

7. Smoker WRK and Khanna G. Imaging the craniocervical junction. *Childs Nerv Syst.* 2008 Oct; 24 (10): 1123-1145
8. Brito JNPO, Santos BAD, Nascimento IF, Martins LA, Tavares CB. Basilar invagination associated with chiari malformation type I: A literature review. *Clinics (Sao Paulo).* 2019;74:e653. doi:10.6061/clinics/2019/e653
9. Dantas FLR, Dantas F, Caires AC, Botelho RV. Natural History and Conservative Treatment Options in Chiari Malformation Type I in Adults: A Literature Update. *Cureus.* 2020;12(12):e12050. Published 2020 Dec 13. doi:10.7759/cureus.12050
10. Holly LT, Batzdorf U. Chiari malformation and syringomyelia. *J Neurosurg Spine.* 2019;31(5):619-628. doi:10.3171/2019.7.SPINE181139
11. Pinter NK, McVige J, Mechtler L. Basilar Invagination, Basilar Impression, and Platybasia: Clinical and Imaging Aspects. *Curr Pain Headache Rep.* 2016;20(8):49. doi:10.1007/s11916-016-0580-x
12. Rosenblum JS, Pomeranec IJ, Heiss JD. Chiari Malformation (Update on Diagnosis and Treatment). *Neurol Clin.* 2022;40(2):297-307. doi:10.1016/j.ncl.2021.11.007
13. Expert Panel on Pediatric Imaging, Radhakrishnan R, Shea LAG, et al. ACR Appropriateness Criteria® Ataxia-Child. *J Am Coll Radiol.* 2022;19(11S):S240-S255. doi:10.1016/j.jacr.2022.09.010
14. Expert Panel on Neurologic Imaging, Pallavi S, Utukuri MD, et al. ACR Appropriateness Criteria® Headache. Available at <https://acsearch.acr.org/docs/69482/Narrative/> American College of Radiology. 2022.
15. Expert Panel on Pediatric Imaging:, Hayes LL, Palasis S, et al. ACR Appropriateness Criteria® Headache-Child. *J Am Coll Radiol.* 2018;15(5S):S78-S90. doi:10.1016/j.jacr.2018.03.017