

## Patient Report

# Type I Chiari malformation presenting with laryngomalacia and dysphagia

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**Abstract** We describe clinical features of an infant with laryngomalacia and dysphagia caused by type I Chiari malformation (CM-I). A 12-month-old child presented with a 6 month history of progressive stridor, dysphagia, and gastroesophageal reflux. Examination of the airway and swallowing function indicated mild laryngomalacia and aspiration with all consistencies. Magnetic resonance imaging of the brain indicated CM-I. Symptoms were resolved after posterior fossa decompression. CM-I, typically diagnosed later in life, should be considered in the differential diagnosis of laryngomalacia and dysphagia. High clinical suspicion and thorough search for abnormalities ensure early diagnosis and proper management of children with neurologic variant laryngomalacia.

**Key words** dysphagia, infant, laryngomalacia, stridor, type I Chiari malformation.

Laryngomalacia, characterized by collapse of supraglottic structures during inspiration, is the most common congenital abnormality of the larynx.<sup>1–3</sup> It is estimated to account for 60–75% of congenital stridor cases. Patients with laryngomalacia present with high-pitched, inspiratory stridor that worsens during crying or in supine position. Stridor progressively worsens by 6 months of age. Laryngomalacia resolves without surgery in up to 90% of patients during the second year of life. Development of failure to thrive, feeding difficulties, upper airway obstruction, and severe dyspnea warrant surgical intervention. Poor surgical outcome has been documented in the presence of neurological impairment, cardiac abnormalities, medical comorbidities, congenital anomalies, or synchronous airway lesions.

Rare forms of laryngomalacia include state-dependent laryngomalacia and neurologic variant laryngomalacia.<sup>4–8</sup> State dependent laryngomalacia is characterized by late-onset laryngomalacia associated with stridor or upper airway obstruction induced by exercise or sleep. Neurologic variant of laryngomalacia has been documented in children with neurologic abnormalities such as type I Chiari malformation (CM-I), and C1 posterior arch compression of the cervicomedullary junction. To date, laryngomalacia caused by CM-I has been documented in two children.<sup>7,8</sup> Delayed diagnosis of neurologic variant of laryngomalacia resulted in failure of supraglottoplasty.<sup>7</sup> Better understanding of the typical presentation, and clinical course of rare forms of laryngomalacia, potentially improves diagnosis and management of these children. We describe the diagnosis and management of an infant with laryngomalacia and dysphagia caused by CM-I and emphasize both clinical and radiological findings of this rare condition.

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## Case report

A 12-month-old Caucasian girl presented with a 6 month history of progressive stridor and dysphagia. The patient was born at full term without complications. She began to cough and choke on feeding, and have stridor at night without an inciting event at 6 months of age. Coughing and choking with feeding were associated with both solid and liquid food. The stridor was intermittent and high-pitched in nature. There was no reported history of apnea, pauses in breathing, or cyanosis. The patient was diagnosed with gastroesophageal reflux disease and was placed on acid suppressive treatment (i.e. lansoprazole). The loudness and frequency of stridor progressively worsened. The patient started to have stridor during the day and developed suprasternal retractions. She was also treated for bronchiolitis, croup, and asthma. Despite medical management, the stridor and dysphagia progressively worsened, and she was referred to hospital for further management.

On examination, she had intermittent, high-pitched inspiratory stridor. Neurological exam showed hyperextension of the head, normal muscle tone and no focal deficit. She was alert and had normal coordination. She sat up on her own and was able to stand with holding. She had not taken her first step. Flexible laryngoscopy showed intact bilateral vocal fold motion, slightly foreshortened aryepiglottic folds, and mild arytenoid edema without inward movement of the arytenoids. She was taken to the operating room for direct laryngoscopy and bronchoscopy that showed omega-shaped epiglottis, slightly foreshortened aryepiglottic fold, mild arytenoid edema, and slight, non-obstructive compression of the anterior tracheal wall suggestive of aberrant innominate artery. Videofluoroscopic swallow demonstrated pooling in the valleculae, premature spillage to the pyriform sinuses and aspiration of all liquid consistencies. Due to the constellation of late-onset laryngomalacia symptoms, dysphagia, signs of mild laryngomalacia, and failure to improve with medical treatment, possible neurologic etiology was considered. Magnetic resonance imaging (MRI) of the neck and chest was

normal with no aberrant innominate artery. MRI of the brain showed CM-I with the cerebellar tonsils protruding to C2 level (Fig. 1). There was syrinx within the spinal cord extending from C2 through to C6. The child underwent posterior fossa decompression with complete resolution of the respiratory symptoms and dysphagia at 6 month follow up.

## Discussion

Type I Chiari malformation is a brainstem abnormality characterized by inferior displacement of the cerebellar tonsils through the foramen magnum. MRI is the gold standard for diagnosis of CM-I, which is identified by the herniation of one tonsil below the foramen magnum by  $>5$  mm or both tonsils by 3 mm. Earlier studies have documented CM-I in adults primarily, but, with the advent of MRI, this condition has been increasingly diagnosed in children. The incidence of CM-I in children ranges from 0.6% to 3.6%.<sup>9,10</sup> Patients with this condition can be asymptomatic but typically present with persistent headaches. Occasionally lower cranial nerve deficits are present. Sensory and motor deficits may occur if a syrinx is present.

Diagnosis of CM-I may be delayed in infants and very young children due to the difficulty of eliciting a history consistent with this condition; atypical symptoms; and clinical presentation mimicking other non-neurologic conditions seen in a routine pediatric otolaryngology practice. Infants cannot verbalize their symptoms and headache may manifest as mere irritability and crying. Lower cranial nerve dysfunction that manifests as recurrent aspiration, oropharyngeal dysphagia, and stridor may be present in up to 78% of children under 3 years old with CM-I. The clinical picture of CM-I in infants may be mistaken for laryngomalacia and/or gastroesophageal reflux disease, which are more common diagnoses in

infants. Earlier studies have documented CM-I and laryngomalacia in two patients who were symptomatic before 5 months of age. The present patient had laryngomalacia associated with late-onset stridor, dysphagia, and gastroesophageal reflux after 6 months of age. Presence of these symptoms does not always necessitate a work-up for neurologic disease; hence, diagnosis and treatment of neurologic variant laryngomalacia may be delayed. Furthermore, patients may fail to improve after multiple supraglottoplasties.<sup>7</sup> Due to discordance between clinical history and examination findings in the present case, MRI of the brain was done to delineate underlying neurological abnormality. Identification of CM-I and resolution of symptoms after neurosurgical intervention supported the diagnosis of neurologic variant laryngomalacia. In agreement with previous clinical reports,<sup>7,8</sup> the present findings emphasize the crucial role of early identification of symptomatic CM-I in surgical planning for children with neurologic variant laryngomalacia because neurosurgical interventions such as posterior fossa decompression, duraplasty, and shunting can resolve symptoms.

## Conclusion

Type I Chiari malformation, although typically diagnosed later in life, should be considered in the differential diagnosis of laryngomalacia and dysphagia. Combination of high clinical suspicion and thorough search for abnormalities in other systems ensures early diagnosis, proper management, and prevention of complications in infants with neurologic variant laryngomalacia.

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## References

- 1 Valera FC, Tamashiro E, de Araújo MM, Sander HH, Küpper DS. Evaluation of the efficacy of supraglottoplasty in obstructive sleep apnea syndrome associated with severe laryngomalacia. *Arch. Otolaryngol. Head Neck Surg.* 2006; **132**: 489–93.
- 2 Day KE, Discolo CM, Meier JD, Wolf BJ, Halstead LA, White DR. Risk factors for supraglottoplasty failure. *Otolaryngol. Head Neck Surg.* 2012; **146**: 298–301.
- 3 Denoyelle F, Mondain M, Gresillon N, Roger G, Chaudre F, Garabedian EN. Failures and complications of supraglottoplasty in children. *Arch. Otolaryngol. Head Neck Surg.* 2003; **129**: 1077–80.
- 4 Oomen KP, Modi VK. Occult laryngomalacia resulting in obstructive sleep apnea in an infant. *Int. J. Pediatr. Otorhinolaryngol.* 2013; **77**: 1617–19.
- 5 Richter GT, Rutter MJ, DeAlarcon A, Orvidas LJ, Thompson DM. Late-onset laryngomalacia: A variant of disease. *Arch. Otolaryngol. Head Neck Surg.* 2008; **134**: 75–80.
- 6 Chan DK, Truong MT, Koltai PJ. Supraglottoplasty for occult laryngomalacia to improve obstructive sleep apnea syndrome. *Arch. Otolaryngol. Head Neck Surg.* 2012; **138**: 50–54.
- 7 Petersson RS, Wetjen NM, Thompson DM. Neurologic variant laryngomalacia associated with Chiari malformation and cervicomedullary compression: Case reports. *Ann. Otol. Rhinol. Laryngol.* 2011; **120**: 99–103.



**Fig. 1** Sagittal T2-weighted magnetic resonance imaging of the cervical spine showing herniation of the cerebellar tonsils below the foramen magnum (arrow).

- 8 Portier F, Marianowski R, Morisseau-Durand MP, Zerah M, Manac'h Y. Respiratory obstruction as a sign of brainstem dysfunction in infants with Chiari malformations. *Int. J. Pediatr. Otorhinolaryngol.* 2001; **57**: 195–202.
- 9 Meadows J, Kraut M, Guarnieri M, Haroun RI, Carson BS. Asymptomatic Chiari type I malformations identified on magnetic resonance imaging. *J. Neurosurg.* 2000; **92**: 920–26.
- 10 Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO. Chiari malformation type I and syrinx in children undergoing magnetic resonance imaging. *J. Neurosurg. Pediatr.* 2011; **8**: 205–13.