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SLEEP MEDICINE PEARLS

Arnold-Chiari malformation: an uncommon etiology for a brief resolved, unexplained event in an infant

Nancy Joseph, MD[1,2]; Dhanashree Rajderkar, MD[3]; Dawn Baker, APRN[2,4]; Sreekala Prabhakaran, MD[2,4]

[1]Division of Hospital Medicine, University of Florida, Gainesville, Florida; [2]Department of Pediatrics, University of Florida, Gainesville, Florida; [3]Department of Radiology, University of Florida, Gainesville, Florida; [4]Division of Pulmonology, University of Florida, Gainesville, Florida

A 57-day-old male infant was referred to the sleep clinic for evaluation of episodes of apnea. The infant was born at term with a birth weight of 7 lb 7 oz. The mother reported that since birth the infant had intermittent episodes of apnea along with inconsolable crying. After one of these episodes, when he was aged 31 days, the infant was taken to the emergency department and admitted for overnight observation. Baseline electrocardiogram was normal, and no seizure-like activity was noted. During physical examination, signs of an arching back and inconsolable crying were noted. Gastroesophageal reflux was suspected, ranitidine was prescribed, and he was referred for further workup. At the clinic, he had normal vital signs, weight was in the 90th percentile, and he was alert and tracking well. He had no focal neurologic deficits and no pathological signs. The patient showed no improvement in apneic episodes after treatment.

QUESTION: What should be the next step in the evaluation of this patient?

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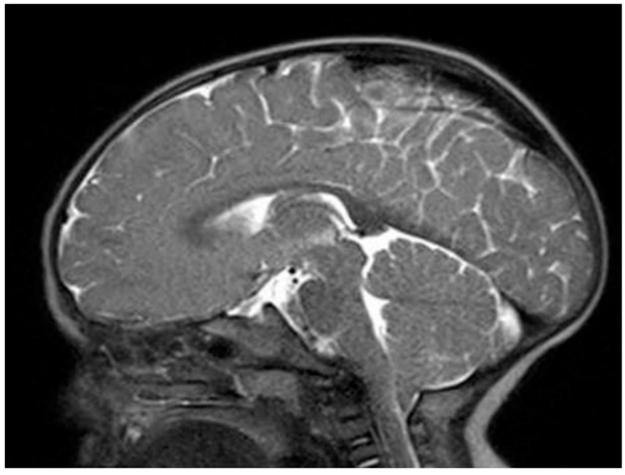
ANSWER: Polysomnography

DISCUSSION

The polysomnography showed a sleep efficiency of 84%, with four obstructive apneas, 56 central apneas, and nine hypopneas. The lowest saturation was 86%, with saturation less than 93% for around 7% of the sleep time. The total apnea-hypopnea index was 9.5 events/h (7.7 from central sleep apnea and 1.8 from obstructive sleep apnea). The central sleep apneas were 10–12 seconds long and commonly were not postarousal central apneas. There was no periodic breathing. The transcutaneous CO_2 > 7 torr above baseline was < 5% of total sleep time. Supplemental oxygen at 100 ml resolved the desaturations, but central sleep apneas persisted. Magnetic resonance imaging of the brain and brainstem was obtained and revealed low-lying cerebellar tonsils about 8 mm below the foramen magnum, consistent with Arnold-Chiari type 1 malformation **(Figure 1).** Suboccipital craniectomy was performed with cervical laminectomy. Postoperative sleep study showed apnea-hypopnea index of 2.3 events/h and central sleep apnea index of 0.9 events/h without any significant desaturations.

This patient's initial presentation warrants the diagnosis of brief resolved, unexplained event. It is important to stratify whether the patient is in the low-risk or high-risk category before testing is initiated per American Academy of Pediatrics Guidelines.[1] The high-risk brief resolved, unexplained event category includes age younger than 60 days, born at < 32 weeks' gestation and corrected gestational age < 45 weeks, cardiopulmonary resuscitation administered more than 1 minute and not first event. Etiology includes gastroesophageal reflux, seizures, respiratory infection or cardiac arrhythmia, and structural malformations of the brain. History, physical examination, and structured evaluation are key to identifying the etiology.

Figure 1—Low-lying cerebellar tonsils about 8 mm below the foramen magnum, consistent with Arnold-Chiari malformation.



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1. Arnold-Chiari type 1 malformation is a congenital brainstem abnormality characterized by caudal herniation of cerebellar tonsils through the foramen magnum resulting in crowding at the craniocervical junction.[2]
2. It has been postulated that central apnea may occur because of compression of the respiratory center, dysfunction of the ascending medullary activating system, abnormal chemosensitivity, and paralysis of movement in the upper respiratory wall.[3]
3. Magnetic resonance imaging of the brain is a useful adjunct in evaluation of central apnea in infants[4]
4. Surgical decompression is the proposed treatment, and the effect of surgery differs among patients based on the use of posterior fossa decompression with dural scoring vs duraplasty.[5,6]

CITATION

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Address correspondence to: Nancy Joseph, MD, Division of Hospital Medicine, Department of Pediatrics University of Florida, 1600 SW Archer road, PO Box 100297 UFHSC. Gainesville, FL 32610; Email: najoseph@ufl.edu

DISCLOSURE STATEMENT

All authors have seen and approved the manuscript. Work for this study was performed at the University of Florida. None of the authors has any financial disclosures or conflict of interest.

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