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Case Reports

Cheyne-Stokes Respiration in a 17-Year-Old Boy Awaiting Heart Transplantation

Nooralam A. Rai, MD[1]; Aliva De, MD[1]; Carin Lamm, MD[1]

[1]Department of Pediatrics, New York-Presbyterian Columbia University Medical Center, New York, New York

Cheyne-Stokes respiration is a pattern of alternating central apnea and hyperpnea. It is well described in adults with congestive heart failure, but not in children.

We report the case of a 17-year-old boy whose systolic heart failure was complicated by Cheyne-Stokes respiration. He was given supportive therapy until heart transplant, after which his Cheyne-Stokes respiration clinically resolved. Clinicians should be aware of this uncommon condition in pediatric and adolescent patients who have advanced heart failure and irregular breathing. ***(Tex Heart Inst J 2021;48(4):e207345)***

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Corresponding author:
Nooralam A. Rai, MD, Department of Pediatrics, CHC 701, New York-Presbyterian Columbia University Medical Center, 3959 Broadway, New York, NY 10032

E-mail:
nr2640@cumc.columbia.edu

Congestive heart failure (HF) may be accompanied by nocturnal oxygen desaturation caused by obstructive sleep apnea (SA), central SA with Cheyne-Stokes respiration (CSR), or sleep-related hypoventilation. Cheyne-Stokes respiration, an abnormal breathing pattern, is present in up to 40% of adults with congestive HF[1,2]; however, it is infrequently reported in children[3] and thus may be underrecognized. A history of irregular breathing or frequent apneas in a child with reduced cardiac function warrants polysomnographic investigation, because undetected CSR can lead to worsened outcomes just as in adult patients.[4] Our report highlights the importance of recognizing and treating this complication of HF in children and indicates a need for further studies of sleep-disordered breathing in this population.

Case Report

A 17-year-old boy who had hereditary dilated cardiomyopathy was hospitalized for worsening HF and was awaiting a heart transplant. Oxygen desaturation was detected when he slept. Frequent pauses in breathing not associated with snoring were followed by multiple deep breaths, and intermittent oxygen desaturation as low as 80% on room air. The patient had clear breath sounds bilaterally, a grade 2/6 systolic ejection murmur, and no peripheral edema. An echocardiogram showed decreased left ventricular (LV) function with an ejection fraction (EF) of 34%. When awake and breathing room air, his oxygen saturation was 96% on finger oximetry.

A polysomnogram revealed an apnea-hypopnea index of 13.5 events/hr and a central apnea index of 12.6/hr. Beginning 3 minutes after sleep onset, multiple central apneas, separated by crescendo and decrescendo changes in breathing amplitude consistent with CSR, were detected during stages 3 and 4 of nonrapid eye movement (NREM) sleep (Fig. 1). These episodes lasted 52 minutes. A second period of CSR in the early morning, associated with stage 2 NREM sleep, lasted 27 minutes. This pattern was not detected during rapid eye movement (REM) sleep. The CSR persisted for 21% of total sleep time. The periods of oxygen desaturation were associated with central apneas. The patient's lowest oxygen saturation was 79%, and it was <90% for 35 minutes of total sleep time. His highest end-tidal CO_2 was 41 mmHg (average, 35 mmHg).

The patient could not tolerate continuous positive airway pressure (CPAP) and removed the nasal mask soon after its placement. Accordingly, supplemental oxygen was provided at a flow rate of 2 L/min through a nasal cannula. Adaptive servoventilation was not implemented because of its association with increased mortality rates in adults with symptomatic chronic HF and LVEF ≤45%.[5] During oxygen therapy, fewer

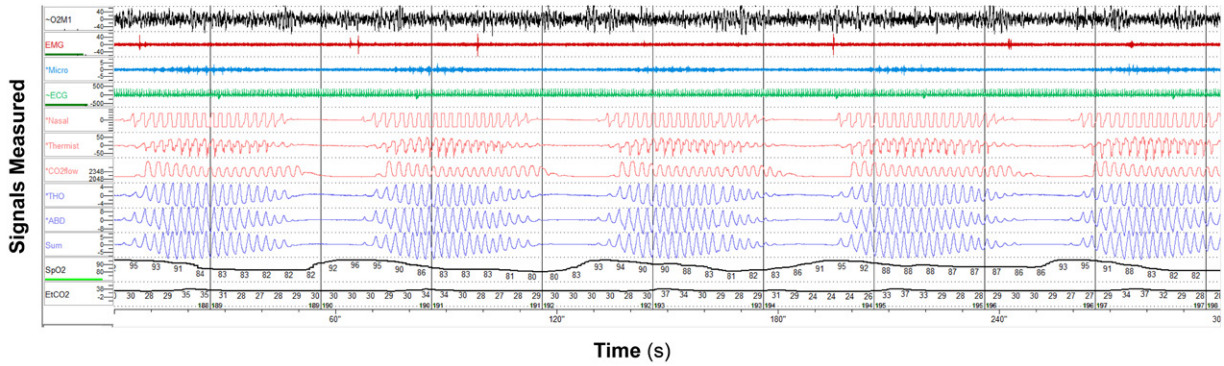

Fig. 1 *Polysomnogram shows the patient's episodes of central apnea between crescendo and decrescendo changes in tidal breathing, consistent with Cheyne-Stokes respiration.*

ABD = abdominal movement measured by inductance plethysmography; CO2flow = air flow measured by end-tidal CO_2; ECG = electrocardiogram; EMG = chin electromyogram; EtCO2 = end-tidal CO_2 (mmHg); Micro = snoring microphone; Nasal = nasal-pressure cannula measuring airflow; O2M1 = occipital electroencephalogram; SpO2 = oxygen saturation measured by pulse oximetry; Sum = sum of abdominal and thoracic movements; Thermist = airflow measured by thermistor; THO = thoracic movement measured by inductance plethysmography

apnea episodes were witnessed and no desaturation was recorded. One week after the polysomnogram, the patient underwent heart transplant; he was placed on CPAP therapy for 2 days and was then removed from respiratory support. He was monitored for another week in the hospital, and no episodes of irregular breathing or oxygen desaturation were observed.

Three months after transplant, an echocardiogram showed normal biventricular systolic function and an LVEF of 55%. A repeat sleep study to ensure improvement in SA was not possible because of the family's geographic and social constraints; however, the patient's parents reported that he was sleeping well without pauses in breathing.

Discussion

Cheyne-Stokes respiration is a type of periodic breathing characterized by central apnea alternating with hyperpnea in a crescendo-decrescendo pattern of changing tidal volumes. It occurs more frequently during NREM sleep than during wakefulness or REM sleep. The probable multifactorial mechanism involves increased chemoreceptor sensitivity with an altered apnea threshold that leads to periodic hyperventilation followed by apnea. It has been well described in adults with congestive HF, but not in children. A prospective observational study by den Boer and colleagues[6] in children with HF showed that CSR was detected in 8% of cases, all in patients ≥12 years of age. The authors proposed that central SA, which includes CSR, may occur less often in children because one cause is nocturnal fluid shift, wherein fluid accumulates in the legs during the day, then stimulates the pulmonary irritant receptors in recumbent posture, triggering hyperventilation. Peripheral edema is not often seen in children with HF, which may partly explain the low incidence of central SA in the pediatric population.[6] Polysomnographic diagnosis is advantageous because central SA is an independent risk factor for reduced survival in adult patients with HF.[7] Among adults with systolic HF and central SA, those who had more characteristics of CSR had a greater number of adverse events, such as hospitalization and death from worsening HF.[8] Less is known about the significance of CSR in young children with HF; Combs and colleagues[9] found that sleep-disordered breathing, particularly central SA, was associated with worse outcomes in hospitalized infants who had congenital heart disease.

Untreated central SA, including CSR, is thought to worsen cardiac function through several mechanisms, including cyclical sympathetic nervous system activation, generation of reactive oxygen species, systemic inflammation, and worsened endothelial dysfunction,[7] and it can contribute to pulmonary hypertension.[10] Thus, although the mainstay of treatment is optimizing HF management, targeted therapy involving positive airway pressure or supplemental oxygen[11] should be considered to avoid the pathophysiologic consequences of persistent CSR. In addition, CPAP can reduce LV afterload and improve LVEF in patients who have CSR and HF.[1] The resolution of CSR after heart transplant has been reported.[3] Although we could not perform a repeat sleep study in our patient, his parents' report indicated that he had recovered.

Our report serves to remind cardiologists, pulmonologists, and pediatricians that CSR is an important sleep-related manifestation of congestive HF. Further knowledge is needed regarding the clinical significance of CSR in pediatric patients and the sleep-related outcomes after heart transplant in this population.

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