CASE REPORTS

Absence of Device-Device Interaction (DDI) in a Patient with Cardiac and Diaphragmatic Pacemakers for Congenital Central Hypoventilation Syndrome

MOHAMMAD-REZA MOVAHED,* MEHRDAD JALILI,†, and NAFIZ KICIMAN‡

From the *Department of Medicine, Division of Cardiology, †Department of Pediatrics, and ‡Division of Pediatric Intensive Care and Division of Pediatric Cardiology, University of California, Irvine, California

MOVAHED, M.R., ET AL.: Absence of Device-Device Interaction (DDI) in a Patient with Cardiac and Diaphragmatic Pacemakers for Congenital Central Hypoventilation Syndrome. *Autonomic control of ventilation is impaired in patients with Ondine's curse or congenital central hypoventilation syndrome (CCHS), but voluntary control remains intact. Bradyarrhythmias can be life threatening. In a patient with CCHS and long sinus pause requiring cardiac pacemaker insertion, a diaphragmatic pacemaker inserted in early childhood caused diaphragmatic pacer spikes observed during the interrogation of the cardiac pacemaker. Diaphragmatic pacing did not interfere with the cardiac pacemaker function. (PACE 2005; 28:1238–1239)*

bradyarrhythmias*, *arrhythmias*, *Ondine's curse*, *congenital central hypoventilation syndrome (CCHS)*, *cardiac abnormalities*, *syncope*, *pacemaker

Background

Congenital central hypoventilation syndrome (CCHS) is a heterogeneous disorder with impaired autonomic control of ventilation. These patients usually require mechanical ventilation soon after birth, and some patients respond to diaphragmatic pacing. There are many other diseases associated with this syndrome including life-threatening bradyarrhythmias. We report a case of CCHS presenting with sinus node dysfunction.

Case Report

The patient, a 14-year-old man, was born with the diagnosis of CCHS. He has been under our care on chronic ventilatory support since birth, as well as general medical care. He is developmentally delayed and has a seizure disorder. Soon after birth, he was diagnosed with Hirshsprung's disease which required surgical repair. He required home ventilatory support 24 hours a day during the first few years of life, with a need for daytime ventilatory support that decreased as the child grew. He underwent placement of a diaphragmatic pacer in order to be completely independent during the day. He was noted to have prolonged sinus pauses (Fig. 1) with heart rates recorded in the low 40s and high 30s. A dual chamber pacemaker was inserted. During pacemaker interrogation, diaphragmatic pacing spikes were noted on the intracardiac ECG as high frequency oscillations every 3 seconds (Fig. 2). Despite this interference, cardiac pacemaker function was not affected. He has now been followed for 18 months since the cardiac pacer was inserted, and is doing well and remains asymptomatic.

Discussion

Patients with CCHS have impaired ventilatory response to hypoxia or hyperapnea, usually during sleep.[1–3] The CCHS is thought to be secondary to the insensitivity of the central chemoreceptors to carbon dioxide. Patients usually require mechanical ventilation soon after birth. Diaphragmatic pacing has been effective in the improvement of symptoms in some patients.[4–8] The cause of the long sinus pause in our patient was not certain. Based on the presence of wandering atrial pacemaker vs junctional rhythm before the onset of long sinus pause, it suggests vasovagal mechanism. Autonomic dysfunction also affects cardiac rhythm. Bradyarrhythmias, vasovagal symptoms, and asystole have been reported in patients with CCHS, which may require pacemaker implantation.[9–13]

Sleep-related disorder of ventilation, the so-called obstructive or central sleep apnea, can occur in children and adults, and resembles CCHS, which cause hypoxia during sleep. However, these sleep disorders are not congenital, and are mostly secondary to airway obstruction during sleep or central apnea secondary to concomitant disorders involving heart or central nervous system such as

Address for reprints: Mohammad-Reza Movahed, M.D., Ph.D., F.A.C.P., F.A.C.C., F.S.C.A.I., F.C.C.P., Assistant Clinical Professor, Director of Nuclear Cardiology, University of California, Irvine Medical Center, 101 The City Drive, Bldg 53, Rm 100, Orange, CA 92868-4080. Fax: (714) 456 8895; e-mail: mmovahed@uci.edu

Received May 20, 2005; revised July 7, 2005; accepted July 13, 2005.

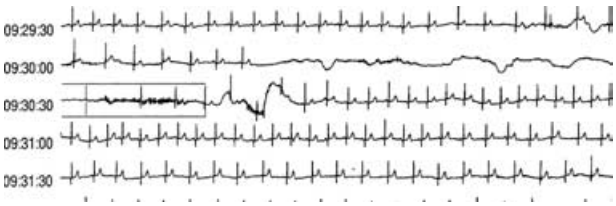

Figure 1. *Long sinus pause over 3 seconds is seen during 24-hour Holter monitor recording.*

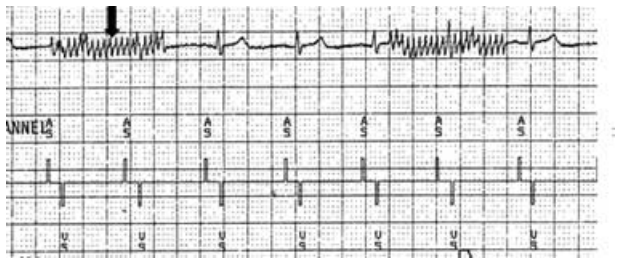

Figure 2. *Intracardiac pacemaker recording: Diaphragmatic pacing spikes are seen as high frequency oscillations after every three beats (arrow) together with intracardiac electrocardiagram during cardiac pacemaker interrogation (Type: Medronic, Kappa KDR921, atrial pacing threshold: 0.5 v/0.52 ms, ventricular pacing threshold: 0.25 v/0.52 ms, P wave: 1.4 mv, R wave: 5.6 mv, atrial sensitivity setting: 0.5 mv, ventricular sensitivity setting: 2.0 mv).*

prematurity. Central sleep apnea (CSA) is an acquired sleep disorder that usually occurs in adults and is not a single disease but present as the final pathway in a large group of heterogeneous disorders. It is characterized by apneic events during sleep with no associated ventilatory effort.[14]

In a survey of 32 patients with CCHS, heart block and sick sinus syndrome occurred in two patients, requiring permanent cardiac pacemaker insertion.[12] In a larger survey of 196 patients with CCHS, cardiac pacing was required in 4.1% of the patients.[13]

Dual chamber pacemaker insertion with rate drop response function can significantly reduce the syncopal episodes in patients with vasovagal syncope and should be programmed in patients suspected of this disorder.[15,16]

Our patient had concomitant diaphragmatic pacing. Interestingly, during follow-up cardiac pacemaker interrogation, simultaneous diaphragmatic pacing spike was recorded on the intracardiac ECG, without any effect on the cardiac pacemaker function.

In order to prevent sudden death, we suggest comprehensive evaluation of patients with CCHS for bradyarrhythmias.

Conclusion

Autonomic dysfunction is common in patients with CCHS, leading to pulmonary and cardiac rhythm abnormalities. Bradyarrhythmias are a life-threatening complication of autonomic dysfunction. Therefore, comprehensive evaluation of patients with CCHS is recommended for early diagnosis and appropriate treatment of bradyarrhythmias.

References

1. Idiopathic congenital central hypoventilation syndrome: Diagnosis and management. American Thoracic Society. Am J Respir Crit Care Med 160(1):368–373. Review.
2. Gozal D, Harper RM. Novel insights into congenital hypoventilation syndrome. Curr Opin Pulm Med 1999; 5(6):335–338.
3. Marcus CL, Bautista DB, Amihyia A, Ward SL, Keens TG. Hypercapnic arousal responses in children with congenital central hypoventilation syndrome. Pediatrics 1991; 8(5):993–998.
4. Flageole H. Central hypoventilation and diaphragmatic eventration: Diagnosis and management. Semin Pediatr Surg 2003; 12(1):38–45.
5. Fitzgerald D, Davis GM, Gottesman R, Fecteau A, Guttman F. Diaphragmatic pacemaker failure in congenital central hypoventilation syndrome: A tale of two twiddlers. Pediatr Pulmonol 1996; 22(5):319–321.
6. Hunt CE, Matalon SV, Thompson TR, Demuth S, Loew JM, Liu HM, Mastri A, Burke B. Central hypoventilation syndrome: Experience with bilateral phrenic nerve pacing in 3 neonates. Am Rev Respir Dis 1978; 118(1):23–28.
7. Weese-Mayer DE, Morrow AS, Brouillette RT, Ilbawi MN, Hunt CE. Diaphragm pacing in infants and children. A life-table analysis of implanted components. Am Rev Respir Dis 1989; 139(4):974–979.
8. Meisner H, Schober JG, Struck E, Lipowski B, Mayser P, Sebening F. Phrenic nerve pacing for the treatment of central hypoventilation syndrome—state of the art and case report. Thorac Cardiovasc Surg 1983; 31(1):21–25.
9. Ogawa T, Kojo M, Fukushima N, Sonoda H, Goto K, Ishiwa S, Ishiguro M. Cardio-respiratory control in an infant with Ondine's curse: A multivariate autoregressive modelling approach. J Auton Nerv Syst 1993; 42(1):41–52.
10. Woo MS, Woo MA, Gozal D, Jansen MT, Keens TG, Harper RM. Heart rate variability in congenital central hypoventilation syndrome. Pediatr Res 1992; 31(3):291–296.
11. Silvestri JM, Hanna BD, Volgman AS, Jones PJ, Barnes SD, Weese-Mayer DE. Cardiac rhythm disturbances among children with idiopathic congenital central hypoventilation syndrome. Pediatr Pulmonol 2000; 29(5):351–358.
12. Weese-Mayer DE, Silvestri JM, Menzies LJ, Morrow-Kenny AS, Hunt CE, Hauptman SA. Congenital central hypoventilation syndrome: Diagnosis, management, and long-term outcome in thirty-two children. J Pediatr 1992; 120(3):381–387.
13. Vanderlaan M, Holbrook CR, Wang M, Tuell A, Gozal D. Epidemiologic survey of 196 patients with congenital central hypoventilation syndrome. Pediatr Pulmonol 2004; 37(3):217–229.
14. Thalhofer S, Dorow P. Central sleep apnea. Respiration 1997; 64(1):2–9. Review.
15. Ammirati F, Colivicchi F, Toscano S, Pandozi C, Laudadio MT, De Seta F, Santini M. DDD pacing with rate drop response function versus DDI with rate hysteresis pacing for cardioinhibitory vasovagal syncope. Pacing Clin Electrophysiol 1998; 21(11 Pt 2):2178–2181.
16. Connolly SJ, Sheldon R, Roberts RS, Gent M. The North American Vasovagal Pacemaker Study (VPS). A randomized trial of permanent cardiac pacing for the prevention of vasovagal syncope. J Am Coll Cardiol 1999; 33(1):16–20.