

Unconventional 2:1 Ventricular Pacing

in a Neonate with Congenital Heart Block and
Biventricular Noncompaction

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Congenital complete heart block with concomitant biventricular noncompaction cardiomyopathy has been reported once previously. Although not universal, when restrictive physiology is present, impaired diastolic filling may pose a distinct challenge to pacing during the neonatal period. We present the case of a neonate with congenital complete heart block and biventricular noncompaction that resulted in severe diastolic dysfunction and atrioventricular dyssynchrony. We intentionally used 2:1 ventricular pacing to provide atrioventricular synchrony with every paced beat, and this resulted in hemodynamic and clinical improvement. This unconventional pacing technique may be beneficial in other neonates who have complete heart block and diastolic dysfunction. (Tex Heart Inst J 2019;46(2):136-8)

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Left ventricular noncompaction (LVNC) is present in 5% to 9% of children who have cardiomyopathy, and up to 20% of these children may have signs of biventricular noncompaction (BVNC).^{1,2} Left ventricular noncompaction presents heterogeneously, manifesting itself as a dilated or hypertrophic phenotype with concordant physiology,^{1,3} which may be more clinically profound in patients with BVNC.² Children with LVNC often have electrocardiographic changes,⁴ but congenital complete heart block (CCHB) in a patient with BVNC has been reported only once previously,⁵ to our knowledge. We present the case of a neonate with a prenatal diagnosis of BVNC and CCHB who needed pacemaker placement shortly after birth, and we discuss our intentional 2:1 pacing strategy to improve the hemodynamic status of this patient with severe diastolic dysfunction.

Case Report

A full-term male infant was born with CCHB, which had been diagnosed by means of fetal echocardiography at 15 weeks' gestation. Maternal test results for anti-Ro/SSA and anti-La/SSB antibodies were negative. A postnatal echocardiogram disclosed severe BVNC with a reduction in internal cavity volumes and ventricular systolic function bilaterally (left ventricular ejection fraction, 0.35). Notably, prominent hepatic and pulmonary vein flow reversals were detected upon each atrial contraction. Electrocardiograms showed multiple P waves per cardiac cycle. Atrial rates after birth were 140 beats/min, and ventricular rates ranged from 50 to 60 beats/min.

The patient was initially treated conservatively, but in response to declining ventricular rates and worsening cardiac output, we placed an Adapta™ ADDR51 dual-chamber epicardial pacemaker (Medtronic) on day of life 4, with use of 4968 CapSure® Epi bipolar steroid-eluting leads (Medtronic) on the atrium and the ventricle. Despite the pacemaker's VVI programming at 80 beats/min, the patient's low-cardiac-output state persisted, necessitating increased inotropic support with use of epinephrine, vasopressin, dopamine, and, eventually, milrinone. An increased pacing rate of 120 beats/min resulted in worsening peripheral perfusion, so the device was ultimately reprogrammed to VVI at 90 beats/min. Given an intrinsic atrial rate exceeding 150 beats/min and the patient's poor tolerance of higher pacing rates, we did not attempt atrioventricular (AV) sequential pacing, which would have further reduced diastolic filling time in the presence of severe diastolic dysfunction and small ventricular cavities.

Considering the patient's substantial diastolic dysfunction, persistent pressor requirement, and lack of AV synchrony, we used an unconventional pacing strategy

Discussion

on day of life 22 to improve his hemodynamic status. To provide some AV synchrony, we programmed the pacemaker to DDD at a lower rate of 80 beats/min and a 2:1 block rate (and upper tracking rate) of 120 beats/min (Fig. 1). This was accomplished by prolonging the postventricular atrial refractory period (PVARP) to 400 ms and the sensed AV delay to 100 ms, thus achieving intentional 2:1 AV block at atrial rates above 120 beats/min. This pacing approach produced AV synchrony with every paced beat, enabling the patient to be weaned from vasopressin, and it reduced mean central venous pressures from 16 mmHg (range, 12–22 mmHg) to 11 mmHg (range, 8–17 mmHg) (Fig. 2).

Because of the patient's persistent heart failure, he was listed at status 1A for ABO-incompatible cardiac transplantation. Medical Genetics was consulted as a part of the pretransplant evaluation because of clinical suspicion of Barth syndrome, which is associated with ventricular noncompaction, substantially elevated levels of 3-methylglutaconic acid and 3-methylglutaric acid on urinalysis, and low total cholesterol (41 mg/dL). The results of a DNA sequence analysis of the taffazin (TAZ) gene were negative for pathogenic mutations of Barth syndrome. Ultimately, the patient underwent successful cardiac transplantation on day of life 28 and remained healthy at outpatient follow-up. At the request of the family, no additional genetic testing was performed after transplantation.

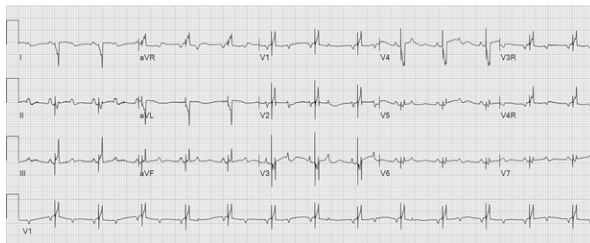


Fig. 1 Twelve-lead electrocardiogram shows 2:1 ventricular pacing with the pacemaker set in DDD mode at 80 beats/min and an upper tracking rate of 120 beats/min. The postventricular atrial refractory period was prolonged to 400 ms, and the sensed atrioventricular (AV) delay was set to 100 ms, resulting in AV synchrony with every paced beat. Nonconducted premature atrial contractions are noted after the 4th, 7th, and 9th ventricular paced beats.

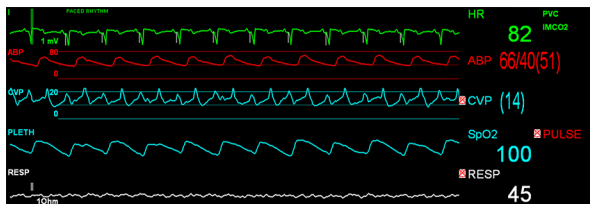


Fig. 2 After atrioventricular synchrony was achieved through 2:1 ventricular pacing, bedside monitor readings show improved central venous pressure (CVP) with each paced beat.

This case highlights distinct management challenges posed by severe diastolic dysfunction in the context of neonatal BVNC and illustrates an alternative pacing technique, which may prove effective in neonates with similar physiology. Left ventricular noncompaction has not classically been associated with CCHB, and we found only one report of a pediatric patient with BVNC and CCHB.⁵ Severe diastolic dysfunction was reportedly not a key physiologic determinant in that patient, nor is it classically the defining feature of patients with LVNC. Epicardial VVI pacing was reportedly used without complication, although details surrounding the pacing approach were not specifically discussed.

Optimal pacing in neonates with CCHB has not been elucidated, and no published consensus guidelines outline pacing nuances in neonates with complex conditions, such as those in our patient. Some experts recommend VVI pacing at a rate limit lower than the intrinsic atrial rate,⁶ whereas others choose to track the atrial rate in a DDD mode.⁷ Glatz and colleagues⁶ presented a case series of 13 high-risk neonates with CCHB who needed pacing within their first 24 hours. The presence of any form of heart disease was a significant predictor of death, and death was universal for those presenting with CCHB and congenital heart disease, except for one patient with spongy myocardium who underwent cardiac transplantation at 67 days of life. Preferred modes of pacing included VVI or VOO for 11 of the 13 patients in this cohort. The authors surmised that atrial tracking and DDD pacing would not be well tolerated in neonates; thus, they preferred to initially pace VVI, increasing the rate slowly over time to achieve physiologic sinus rates.

Of note, in that cohort,⁶ DDD pacing and 1:1 AV conduction attempted in one patient resulted in marked hypotension. The PVARP was lengthened to produce 2:1 AV conduction, which substantially improved the blood pressure. We independently used the same pacing strategy in our neonate who had severe diastolic dysfunction in the context of BVNC. Our patient's hemodynamic instability during the early neonatal period necessitated epicardial pacemaker placement, and a dual-chamber device was chosen because of the perceived future need for AV sequential pacing. Certainly, the restrictive nature of severe BVNC in our patient left him particularly preload-dependent and perhaps more in need of appropriate atrial kick. The 2:1 pacing provided AV synchrony and a more physiologic AV interval with every paced beat and augmented diastolic filling, resulting in improved mean central venous pressures and less inotropic support. The DDD pacing mode with a limited upper tracking rate was not attempted in this patient because of apparent hemodynamic intolerance of higher ventricular rates. However, DDD mode

with a substantially limited upper tracking rate of 80 beats/min may have produced a similar ventricular rate, although with a less physiologic AV interval because of progressive lengthening of the AV interval during pacemaker Wenckebach. Our patient underwent successful heart transplantation 6 days after the initiation of 2:1 pacing, so additional short- and midterm benefits of this pacing are only speculative.

We have presented an unusual case of neonatal BVNC cardiomyopathy associated with severe diastolic dysfunction, complicated by CCHB. Intentionally programming a pacemaker to provide 2:1 AV block is an unconventional pacing technique that may enable intermittent AV synchrony and augment diastolic filling in other neonates who have severe diastolic dysfunction.

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