Pediatr Neonatol 2008;49(6):210–212

EDITORIAL

Myocarditis and Complete Atrioventricular Block: Rare, Rapid Clinical Course and Favorable Prognosis?

Mei-Hwan Wu*

Department of Pediatrics, National Taiwan University Hospital and College of Medicine, National Taiwan University, Taipei, Taiwan

Since the first report in 1947 by Gore and Saphir[1] who described complete atrioventricular block (CAVB) as a rare complication of acute myocarditis, several pediatric studies, mostly case reports, have been reported.[2–26] The pathological mechanisms were attributed to diffuse and severe acute inflammatory changes of the atrioventricular conduction system mainly in the right bundle and the left bundle branches, especially in the terminal portions, focally noted in the AV node and the His bundle.[8] The development of CAVB probably followed the severe acute inflammation of the bundle branches. This may explain why the ventricular rates are most likely associated with wide QRS duration. In addition, there may be a certain degree of lymphocyte infiltration in the myocardium, though to a relatively limited degree as compared to that in patients with compromised myocardial cantractility.[27] In experimental murine models inoculated intraperitoneally with the encephalomyocarditis virus to produce acute myocarditis, CAVB was found in 11% of those with experimental myocarditis: with mononuclear cell infiltrations and necrosis of the AV conduction system in about 27% of those with myocarditis and CAVB and edematous changes in the rest.[28] The severity of the pathological changes of the AV conduction system may reflect the extent of reversibility of the CAVB.

The clinical symptoms of acquired CAVB related to myocarditis are often characterized by acute onset of symptoms of low cardiac output due to a slow ventricular rate that may even lead to hypotension or Stokes-Adams seizures.[2–26] The cardiac enzyme may be abnormal. Electrical instability leading to ventricular tachycardia or ventricular fibrillation may occur. At disease onset, patients may report various specific or non-specific symptoms, such as fever, general malaise, dyspnea, chest pain, abdominal pain, nausea or vomiting. Some of the latest reports emphasized that the gastrointestinal symptoms are often the earliest symptoms of acute myocarditis.[4] Though the CAVB may resolve 1 to 2 weeks after its onset, it may persist. In this issue, Chien et al summarized their experience in nine patients with CAVB presumed to be caused by acute myocarditis.[29] They found that gastrointestinal symptoms, including abdominal pain, nausea or vomiting, was the most common presenting symptoms and was found in 5 out of 9 with CAVB due to acute myocarditis. The mean interval between the first symptoms and the diagnosis (the day when CAVB was confirmed) was 3 days.

Because previous pediatric reports regarding the clinical course of CAVB after myocarditis were mostly case reports or small case series, we hereby summarize the 62 patients (24 male 38 female) who were identified from previous reports and the report by Chien et al in this issue in Table. The age of onset ranged from 13 days to 20 years (mean 2.66 years). Possibly etiology-related pathogens were reported in 18 (29%), including Coxsackie B virus

*Corresponding author. Department of Pediatrics, National Taiwan University Hospital and College of Medicine, National Taiwan University, 8 Chung-Shen South Road, Taipei, Taiwan.
E-mail: wumh@ntu.edu.tw

Table Summary of reported pediatric patients with atrioventricular block due to myocarditis

Reference	No. of cases	Mean age (yr)	Recovery time (days)	Associated tachyarrythmias	Pathogen
Craenen and Moore[10]	1 (M)	6	7	–	–
Thompson and Nolan[11]	1 (F)	9	3	–	Mumps
Johnson and Lee[12]	1 (F)	4	5	–	–
Gould et al[13]	1 (M)	20	2	–	Cox B
Bairan et al[14]	1 (M)	3	P	–	RSV
Schieken and Myers[15]	1 (F)	15	P	–	Cox B
Lim et al[16]	2 (F)	12.5	P	–	–
	5 (F)	15	1		–
Matisonn et al[17]	1 (M)	8	3	–	Diptheria
Giles and Gohd[18]	1 (M)	15	7	–	RSV
Friedli et al[19]	1 (M)	1.5	P	–	Mycoplasma
Reitman et al[20]	1 (F)	4	4	–	EBV
Granath et al[21]	1 (M)	16	5	–	–
Onouchi et al[9]	6 (4M/2F)	5.5	1	–	–
Fujiwara et al[8]	1 (F)	1.4	P	–	–
Liao et al[7]	1 (M)	17	12	–	–
Bromberg et al[22]	1 (F)	3	7	–	–
Mahoney et al[23]	2 (F)	12.5	P	–	Cox B (1 case)
		10	3.5		Cox B (1 case)
Agarwala & Ruschhaupt [24]	1 (M)	12	2	–	Mycoplasma
Rich and McErlean[25]	1 (F)	6	5	–	Varicella
Heusch et al[6]	1 (F)	3	P	–	–
	1 (F)	10	18		–
Khongphatthanayothin et al[26]	2 (F)	10	2	–	–
	6 (F)	2.4	–		
Batra et al[2]	1 (F)	7	5	–	–
Wang et al[4]	1 (F)	0.8	P	VT (3)	Adenovirus (1)
	8 (5M/3F)	7.9	1–10 (3.5)		Mycoplasma (2) Herpers (1)
Heitink-Pollé et al[3]	1(F)	9	2	–	–
Maiers & Ebenroth[5]	1 (M)	20 days	2	JET	
Chien et al[31]	3 (2M/1F)	8.9	P	VT (6)	Mycoplasma (1),
	6 (3M/3F)	9.7	1–12 (6.8)		Cox B (1)

Cox B = Coxsackie B virus; EBV = Ebstein–Barr virus; F = female; M = male; JET = junctional ectopic tachycardia; P = persistent complete atrioventricular block; RSV = respiratory syncytial virus; VT = ventricular tachycardia.

in five, mycoplasma in five and respiratory syncytial virus in two. In the two reports from Taiwan, ventricular tachyarrhythmias occurred in nine (50%) patients.[4,29] Only five (28%) had abnormal cardiac enzyme (CKMB/CK ratio greater than 5%.[4,29] If limited to reports in the last 10 years and to patients with acute onset, permanent AVB was noted in four (19%). Recovery of the AV conduction occurred mostly within the 1st week after the onset.

Therefore, CAVB if occurring as a complication of acute myocarditis most likely manifests as acute onset of low cardiac output due to a slow ventricular rate after a few days of non-specific symptoms. In about half of these cases with acute myocarditis, the rhythms may even progress to life-threatening ventricular tachyarrhythmias. Only about one quarter of them would have abnormal cardiac enzymes. Cardiac support, such as a temporary pacemaker for CAVB, antiarrhythmic agents for ventricular arrhythmias and extracorporeal membranous oxygenation for medically refractory heart failure, may improve the outcome and the prognosis tends to be good. However, significant mortality and morbidity may still occur. Treatment strategies other than acute cardiac support should be explored to optimize the outcome.

References

1. Gore I, Saphir O. Myocarditis: a classification of 1,402 cases. *Am Heart J* 1947;38:827–30.
2. Batra AS, Epstein D, Silka MJ. The clinical course of acquired complete heart block in children with acute myocarditis *Pediatr Cardiol* 2003;24:495–7.

3. Heitink-Pollé KM, Rammeloo L, Hruda J, Plötz FB. Rapid and full recovery after life-threatening complete atrioventricular block: an isolated feature of myocarditis? *Eur J Pediatr* 2004;163:410–11.
4. Wang JN, Tsai YC, Lee WL, Lin CS, Wu JM. Complete atrioventricular block following myocarditis in children. *Pediatr Cardiol* 2002;23:518–21.
5. Maiers JA, Ebenroth ES. Junctional ectopic tachycardia following complete heart block associated with viral myocarditis. *Pediatr Cardiol* 2006;27:367–8.
6. Heusch A, Kühl U, Rammos S, Krogmann ON, Schultheiss HP, Bourgeois M. Complete AV-block in two children with immunohistochemical proven myocarditis. *Eur J Pediatr* 1996; 155:633–6.
7. Liao PK, Seward JB, Hagler DJ, Driscoll DJ. Acute myocarditis associated with transient marked myocardial thickening and complete atrioventricular block. *Clin Cardiol* 1984;7: 356–62.
8. Fujiwara T, Akiyama Y, Narita H, et al. Idiopathic acute myocarditis with complete atrioventricular block in a baby. Clinicopathological study of the atrioventricular conduction system. *Jpn Heart J* 1981;22:275–80.
9. Onouchi Z, Haba S, Kiyosawa N, Shimazu S, Hamaoka K, Kusunoki T. Stokes–Adams attacks due to acute nonspecific myocarditis in childhood. *Jpn Heart J* 1980;21:307–15.
10. Craenen J, Moore D. Stokes–Adams attacks in a small child treated by prolonged use of an indwelling pacing catheter. *Pediatrics* 1976;39:14–7.
11. Thompson WM, Nolan TB. Atrioventricular dissociation associated with Adams–Stokes syndrome presumably due to mumps myocarditis. *J Pediatr* 1966;68:601–7.
12. Johnson JL, Lee LP. Complete atrioventricular heart block secondary to acute myocarditis requiring intracardiac pacing. *J Pediatr* 1971;78:312–6.
13. Gould L, Diaz R, Gomprecht R. Complete heart block in myocarditis—recovery with a pacemaker. *Chest* 1972;62: 230–3.
14. Bairan AC, Cherry JD, Fagan LF, Codd JE. Complete heart block and respiratory syncitial virus. *Am J Dis Child* 1974; 127:264–5.
15. Schieken RM, Myers MG. Complete heart block in viral myocarditis. *J Pediatr* 1975;87:831–42.
16. Lim CH, Toh CS, Chia B, Low L. Stokes–Adams attacks due to acute nonspecific myocarditis. *Am Heart J* 1975;90:172–8.
17. Matisonn RE, Mitha AS, Elliot C. Successful electrical pacing for complete heart block complicating diptheritic myocarditis. *Br Heart J* 1976;38:423–6.
18. Giles TD, Gohd RS. Respiratory syncitial virus and heart disease; a report of two cases. *JAMA* 1976;236:1128–30.
19. Friedli B, Renevey F, Rouge JC. Complete heart block in a young child presumably due to Mycoplasma pneumoniae myocarditis. *Acta Pediatr Scand* 1977;66:385–8.
20. Reitman MJ, Zirin HJ, DeAngelis CJ. Complete heart block in Epstein–Barr myocarditis. *Pediatrics* 1978;62:847–9.
21. Granath A, Kimby E, Sodermark T, Volpe U, Zetterquist S. Stokes–Adams attacks requiring pacemaker treatment in three patients with acute nonspecific myocarditis. *Acta Med Scand* 1980;207:177–81.
22. Bromberg BL, Beekman RH, Heidelberger K. Heart block with viral myocarditis. *J Pediatr* 1986;108:790–830.
23. Mahoney LT, Marvin WJ, Atkins DL, Clark EB, Lauer RM. Pacemaker management for acute onset of heart block in childhood. *J Pediatr* 1985;107:207–11.
24. Agarwala BN, Ruschhaupt DG. Complete heart block from *Mycoplasma pneumoniae* infection. *Pediatr Cardiol* 1991; 12:233–6.
25. Rich R, McErlean M. Complete heart block in a child with varicella. *Am J Emerg Med* 1993;11:602–5.
26. KhongphatthanayothinA, ChotivitayatarakornP, BenjacholamasV, et al. Complete heart block in children at Chulalongkorn Memorial Hospital. *J Med Assoc Thai* 2001;84:S111–7.
27. Morgera T, Di Lenarda A, Dreas L, et al. Electrocardiolography of myocarditis revised: clinical and prognostic significance of electrophysiological changes. *Am Heart J* 1992;124:455–67.
28. Ohmae M, Kishimoto C, Tomioka N. Complete atrioventricular block in experimental murine myocarditis. *J Electrocardiol* 2005;38:230–4.
29. Chien SJ, Liang CD, Lin IC, Huang CF. Myocarditis complicated by complete atrioventricular block: Nine year's experience in a medical center. *Pediatr Neonatal* 2008;49:218–23.