

Association of Heart Block with Uncommon Disease States

Malka Yahalom, MD, DSc, FICA¹ Nathan Roguin, MD² Dante Antonelli, MD¹ Khaled Suleiman, MD¹
Yoav Turgeman, MD^{1,2}

¹ Department of Cardiology, HaEmek Medical Center, Afula, Israel

² Faculty of Medicine, Technion, Haifa, Israel

Address for correspondence Yoav Turgeman, MD, Heart Institute, HaEmek Medical Center, Rabin Avenue 18101, Afula, Israel (e-mail: Yoav_t@clalit.org.il).

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Abstract

A variety of diseases, other than the common Lev–Lenègre disease, are associated with cardiac conduction system abnormalities. These include acute processes, such as acute rheumatic fever, and other disorders, such as sarcoidosis, connective tissue disorders, neoplasms, and bacterial endocarditis with cardiac abscess formation. The purpose of the study is to raise awareness of these rare conditions. We present 10 adult patients (4 males and 6 females) with a mean age of 47 years (range: 19–69), with various rare diseases associated with heart block, who needed temporary or permanent pacemaker therapy in the past two decades. These conditions included acute rheumatic carditis, Wegener granulomatosis, cardiac involvement of metastatic breast cancer, bacterial endocarditis, sarcoidosis, S/P chest radiotherapy, and quadriplegia with syringomyelia postspinal cord injury, and adult congenital heart block. We conclude that patients with these disorders should be followed periodically, to allow for early detection and treatment of cardiac conduction disturbances, with pacemaker therapy.

Keywords

- ▶ heart block
- ▶ association
- ▶ uncommon
- ▶ adults
- ▶ pacemaker
- ▶ prevention
- ▶ follow-up

A variety of diseases, other than common coronary artery disease^{1–3} and Lev–Lenègre syndrome,^{4,5} are associated with cardiac conduction system abnormalities. These include acute processes, such as acute rheumatic fever,^{6,7} and other disorders such as sarcoidosis,^{8,9} Wegener granulomatosis,^{10–13} bacterial endocarditis with cardiac abscess formation,^{14–28} connective tissue disorder,^{29,30} and neoplasms.^{31–36} Cardiac conduction system abnormality has been documented by electrocardiography with calcification of heart valves and valve rings,^{1,37} also following irradiation therapy to the chest,^{38–43} and after spinal cord injury (SCI),^{44–48} as well as after repair of congenital malformations such as ostium primum and tetralogy of Fallot,^{49–51} and in congenital atrioventricular block in adults.⁵²

We describe 10 different cases of adult patients with rare and uncommon disease states associated with heart block, who were treated during the last two decades with pacemaker therapy.

Materials and Methods (I–X)

We present 10 adult patients with various rare diseases associated with heart block, in the last two decades, who needed pacemaker therapy (temporary or permanent) (▶ **Table 1**) (▶ **Figs. 1–3**).

Discussion

The cause of chronic heart block is often obscure when relying solely on clinical grounds.¹ Based on clinical and electrocardiographic evidence, coronary artery disease has been generally accepted by Levine et al² and Friedberg et al³ and others, as the predominant associated disease/cause of heart block.

Other studies suggest that areas of fibrosis involving the conducting system, either alone or in association with scattered areas of fibrosis in the myocardium (Lev–Lenègre syndrome) are responsible for heart block.^{4,5} In an effort to

Table 1 Summary of patients' characteristics and types of heart block

No.	Age	Sex	Clinical presentation		Cardiac conduction abnormalities (ECG)	Associated uncommon disorders	Mode of therapy of cardiac pacing		Figure
			Syncope	Other			Temporary	Permanent	
1	33	F	+	-	CAVB	Recurrent rheumatic carditis	-	+	-
2	19	M	+	-	SB, first degree AV block, AV dissociation	S/P rheumatic carditis	-	+	-
3	43	F	+	-	Intermittent CAVB	Wegener granulomatosis	-	+	1A; 1B
4	68	F	+	-	CAVB	Metastatic breast cancer	+	-	-
5	69	F	+	-	CAVB	Bacterial endocarditis	+	-	-
6	43	M	+	-	CAVB	Sarcoidosis	+	+	2
7	42	M	+	-	CAVB	S/P chest irradiation therapy	+	+	3
8	49	M	-	Fatigue dyspnea	Trifascicular block	Spinal cord injury	-	+	-
9	43	F	-	Fatigue and dizziness	CAVB	Congenital AV block	-	+	-
10	61	F	-	Fatigue and dizziness	CAVB	Mitral annular calcification	-	+	-

Abbreviations: AV, atrioventricular; CAVB, complete AV block; ECG, electrocardiography; F, female; M, male.

improve the clinical diagnosis of the underlying cause of heart block, a retrospective survey was published by Harris et al in 1969, in 65 consecutive patients with chronic heart block who had come to necropsy during 3 years.¹ The etiology and histologic findings of chronic heart block were bilateral bundle-branch fibrosis, cardiomyopathy, coronary artery disease, myocarditis, calcification of valves or valve rings, collagen disease, amyloid deposits, transfusion siderosis, aneurysm of membranous septum, congenital heart block, and syphilitic cardiovascular disease (gumma).¹

Transient advanced atrioventricular conduction block in acute rheumatic fever was described by Zalstein et al,⁶ Malik et al,⁷ Clarke and Keith,⁵³ and by our group.⁵⁴ Other etiological and pathological processes have been attributed to advanced heart block, such as cardiac sarcoidosis,^{8,9} amyloidosis with heart involvement,^{55,56} and cardiac complications of Wegener granulomatosis.¹⁰⁻¹³

There is much evidence of septal involvement of the heart with the formation of abscess in infectious diseases such as acute bacterial endocarditis, involving the heart conduction system.¹⁴⁻²⁸

Collagen diseases and various types of vasculitis have been postulated as a cause of heart conduction disturbances, as described by Bernstein in humans in 1951²⁹ and also in an animal model.³⁰

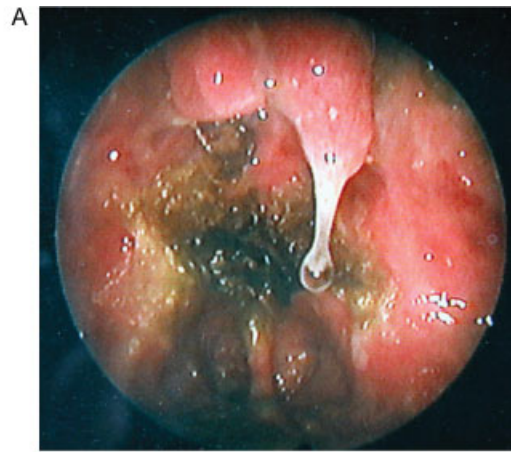
Calcification of heart valves and valve rings has been shown to destroy the cardiac conduction system¹ and is thus associated with advanced heart block.³⁷

Secondary cardiac involvement in metastatic cancer (e.g., breast cancer and lymphoma), especially of the heart conduction system, has been documented.³¹⁻³⁶ The spreading routes are assumed to be via the lymphatic system and blood circulation. In the case of secondary tumors located in the myocardium, it is assumed that the clinical pattern will be proportional to the degree of myocardial infiltration, with a typical presentation of arrhythmic and conduction disturbances with complete atrioventricular blocks, especially where the conduction system has been infiltrated.³²⁻³⁶

The frequency of heart conduction abnormalities following mediastinal irradiation, mainly in patients with Hodgkin disease or breast carcinoma is delayed, and seems to rise with the improvement of therapy and longer survival (of patients).³⁸⁻⁴³ It has been postulated that the younger the patient was when treated with irradiation therapy, the longer the time that has passed since the initial therapy, and the higher the radiation dose given, the more associated cardiac (including heart conduction disturbances) or mediastinal radiation-induced lesions existed.³⁸⁻⁴³

Kaplan et al³⁹ and Cohen et al⁴⁰ noted that when cardiac histopathology was performed in these patients, it disclosed extensive fibrosis of the conduction system and of the atria and ventricles. It was concluded that the severe fibrosis was primarily due to irradiation rather than being secondary to atherosclerotic coronary artery disease.

Another uncommon etiology of heart block has been described in animal models⁴⁴ and humans⁴⁵ suffering from



B Nasal Mucosa Biopsy with Inflammatory and Giant Cells

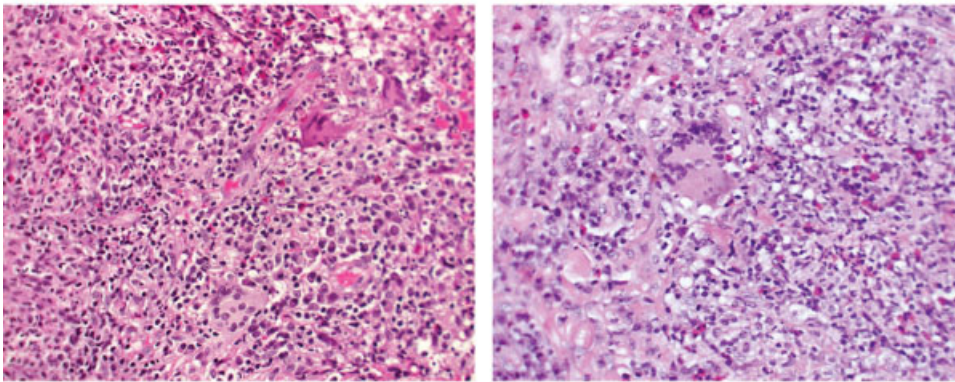


Fig. 1 (A) Cerebrospinal fluid in nasopharynx in Wegener granulomatosis. (B) Nasal mucosa biopsy with inflammatory and giant cells.

SCI, paraplegia, and syringomyelia (cystic degeneration of the spinal cord)—that could be observed months, years, and decades after the primary insult.^{46,47} The mechanisms for the cardiovascular involvement in SCI were attributed to the

sedentary life style and lower daily energy expenditure, combined with a greater prevalence of cardiovascular risk factors and autonomic dysfunction that was documented following the offending event.⁴⁸

Nonnecrotising Granulomas-Sarcoid type

Sarcoid-Granulomas composed of Epithelioid Histiocytes and multinucliated Giant Cells

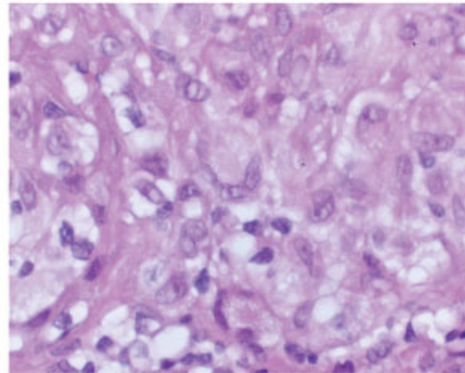
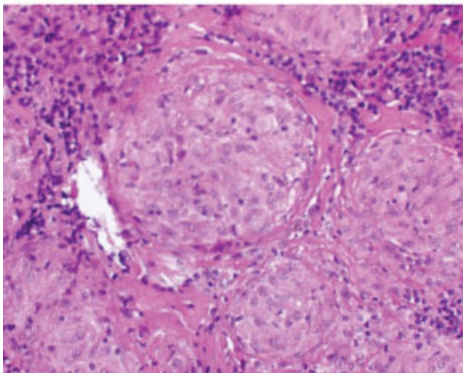


Fig. 2 Lymph node biopsy demonstrating nonnecrotising granulomas-sarcoid type, composed of epithelioid histiocytes and multinucleated giant cells.

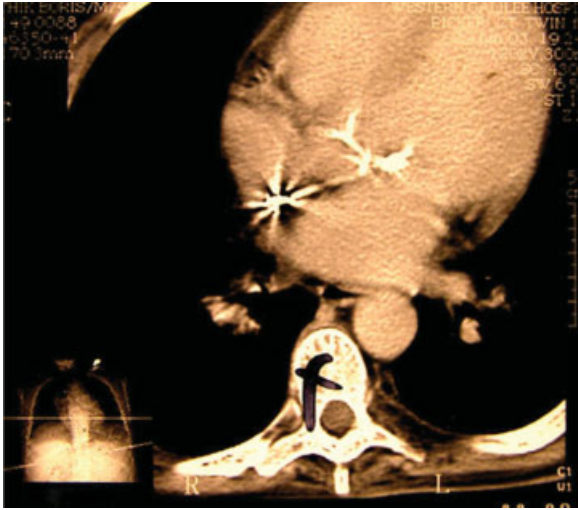


Fig. 3 Computed tomography scan of chest, with evidence of heavy calcification of coronary arteries and mitral annulus calcification.

Miscellaneous etiologies for heart block in adults were: surgical repair of congenital heart disease such as tetralogy of Fallot⁴⁹ and ostium primum defect repair in children⁵⁰; congenital heart block and after transient ischemic attacks⁵¹; congenital complete atrioventricular block in adults⁵² and Chagas heart disease in Latin America.⁵⁷

We have described 10 different cases of adult patients in whom heart block was associated with rare and uncommon diseases, and who were treated during the last two decades with pacemaker therapy.

Conclusions

It is suggested that patients with these disorders should be followed periodically to allow for early detection and timely treatment of cardiac conduction disturbances with pacemaker therapy.

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