SPECIAL ARTICLE

Guideline for Diagnosing Chronic Myocarditis

Japanese Circulation Society (JCS) Task Force Committee
on Chronic Myocarditis#

CHRONIC myocarditis lacks a concise definition[1-12] the term "chronic myocarditis" has been used to describe various conditions of myocarditis[13-15] The Dallas criteria for myocarditis[16,17] do not address this issue. Therefore, in 1991, the Japanese Circulation Society organized a Task Force Committee in order to establish guidelines. The Committee members were all experienced clinical cardiologists and/or cardiac pathologists who were specialists in this field.

This Committee met regularly: in Kurume City (November 1991), Tokyo (November 1992), Ube City (November 1993) and Tokyo (February 1994). Procedures adopted by the Committee included a nationwide survey to isolate suspected cases of chronic myocarditis, and discussions concerning these cases.

A total of 58 cases of chronic myocarditis were suspected, 15 of these were eventually confirmed to be chronic myocarditis. Through discussion and consultation, one case was identified as persistent inflammation type following an acute onset, the other 14 cases were of insidious onset type. Definitions of the two types follow, along with our findings and recommendations.

It is the Committee's hope that these definitions will be applied in both clinical and histopathological recognition of this disease.

Key words:
Chronic myocarditis
Heart failure
Arrhythmias
Active myocarditis
Endomyocardial biopsy
Myocardial scintigram
Autopsy

DEFINITION

Chronic myocarditis consists two types. The first is characterized by prolonged or persistent inflammatory infiltration following an acute onset of myocarditis;* the other is characterized by an insidious onset of myocarditis with chronic processes. The latter may develop heart failure which may mimic dilated cardiomyopathy and/or arrhythmias.

The myocarditis does not include disorders attributed to the following: toxicities (cocaine, amphetamines, etc), chemicals (lithium, arsenic, doxorubicin, etc), physical agents (radiation, heat, etc), allergy, immune disorders such as collagen disease, and sarcoidosis.

REFERENCING MATTERS FOR DIAGNOSIS

1) Observable symptoms and signs suggesting the presence of myocarditis may in-

(Received January 27, 1995; accepted March 9, 1995)

#The Task Force Committee meetings were attended by the following: KYO ADACHI, M.D.(Rapporteur), SHIGEKAZU FUJIOKA, M.D., HISAYOSHI FUJIWARA, M.D., SHINJI FUKUTA, M.D., MICHIAKI HIROE, M.D., SHIGERU ISHIYAMA, M.D., TOHRU IZUMI, M.D., NOBORU KAGAWA, M.D., SACHIO KAWAI, M.D., KEISHIRO KAWAMURA, M.D., YASUSHI KITAURA, M.D., MAKOTO KODAMA, M.D., REIZO KUSUKAWA, M.D., AKIRA MATSUMORI, M.D., MASUNORI MATSUZAKI, M.D., SHIN-ICHIRO MORIMOTO, M.D., TOSHIO NISHIKAWA, M.D., RYOZO OKADA, M.D., TOSHIAKI OKAMOTO, M.D., MORIE SEKIGUCHI, M.D., YOSHINORI SEKO, M.D., SHIN SUZUKI, M.D., HIRONORI TOSHIMA, M.D.(Chairman), HIROSHI YAMAGUCHI, M.D., YOSHIO YAZAKI, M.D. and CHIKAO YUTANI, M.D.

Mailing address: K. Adachi, M.D., Kurume University, School of Medicine, 67 Asahimachi, Kurume 830, Japan

clude heart failure and/or arrhythmias.

2) Endomyocardial biopsy:
   There is accumulation or infiltration** of large and/or small round cells in the myocardium associated with myocytolysis or necrosis of adjacent myocytes. Diffuse interstitial fibrosis, irregular replacement of myocardial fibrosis, and fatty infiltration are also observed. The myocytes show hypertrophy, variation in size, and an irregular arrangement.
3) Myocardial scintigraphy:
   Positive findings obtained through ^{67}Ga-citrate scintigrams, ^{99m}Tc-pyrophosphate scintigrams or ^{111}In-antimyosin antibody scintigrams are valuable indicators of the myocarditis.
4) Autopsy:
   In some cases, endomyocardial biopsy may not show the myocarditis. However, the diagnosis of chronic myocarditis may still be made at autopsy.

*Prolongation or persistence of inflammatory infiltration means that active myocarditis remains present for more than 3 months following the onset of acute myocarditis.
**Cellular infiltration is defined as the existence of more than 5 mononuclear cells per field (magnification ×400), and cellular accumulation is more than 20 mononuclear cells per field (magnification ×400) by optical microscopic examination.

REFERENCES

1. Kelle K: Über primäre chronishe Myocarditis. *Deutsch Arch klin Med* 1892; **49:** 442–456
2. Balchum OJ, McCord MC, Blount SG: The clinical and hemodynamic pattern in nonspecific myocarditis: a comparison with other entities also impairing myocardial efficiency. *Am Heart J* 1956; **52:** 430–443
3. Kline IK, Saphir O: Chronic pernicious myocarditis. *Am Heart J* 1960; **59:** 681–697
4. Kawai S, Shimizu M, Okada R, Ih S: A morphological analysis of chronic myocarditis. *Jpn Circ J* 1987; **51:** 1385–1392
5. Toshima H, Adachi K, Koga Y, Bekki M, Yamamoto K: Two autopsy cases of chronic myocarditis. *In:* Annual Report of Idiopathic Cardiomyopathy Research Committee (Chairman: J. Yasuda): The Ministry of Health and Welfare, the Japanese Government; 1988; 143–145 (in Japanese)
6. Hisaoka T, Kawai S, Ohi H, et al: Two cases of chronic myocarditis mimicking arrhythmogenic right ventricular dysplasia. *Heart Vessels* 1990; **Supple 5:** 51–54
7. Lieberman EB, Hutchins GM, Herskowitz A, Rose NR, Baughman KL: Clinicopathologic description of myocarditis. *J Am Coll Cardiol* 1991; **18:** 1617–1626
8. Morimoto S, Hiramitsu S, Yamada K, et al: Clinical and pathologic features of chronic myocarditis: four autopsy cases presenting as dilated cardiomyopathy in life. *Am J Cardiovasc Pathol* 1992; **4:** 181–191
9. Okabe M, Fukuda K, Nakashima Y, Hiroki T, Arakawa K, Kikuchi M: Lymphocytic active myocarditis characterized by numerous clusters of lymphocytes: a chronic variant of myocarditis? *Am Heart J* 1992; **123:** 128–136
10. Matsumori A: Animal models: pathological findings and therapeutic considerations. *In:* Banatvala JE, editor. Viral infections of the heart. London, Boston, Melbourne, Auckland: Hodder and Stoughton Ltd; 1993: 110–137
11. Kühl U, Strauer BE, Schultheiss H-P: Methylprednisolone in chronic myocarditis. *Postgrad Med J* 1994; **70**(Suppl 1): S35–S42
12. Lauer B, Padberg K, Schultheiss H-P, Strauer B-E: Autoantibodies against human ventricular myosin in sera of patients with acute and chronic myocarditis. *J Am Coll Cardiol* 1994; **23:** 146–153
13. Cecil RL, editor. A Textbook of Medicine, 6th edition. Philadelphia, London: WB Saunders Co; 1944: 1053–1054
14. Price FW, editor. A Textbook of the Practice of Medicine, 8th edition. London, New York, Toronto: Geoffrey Cumberlege, Oxford University Press; 1950; 1022–1024
15. Fenoglio JJ, Ursell PC, Kellogg CF, Drusin RE, Weiss MB: Diagnosis and classification of myocarditis by endomyocardial biopsy. *N Engl J Med* 1983; **308:** 12–18
16. Aretz HT, Billingham ME, Edwards WD, et al: Myocarditis: a histopathological definition and classification. *Am J Cardiovasc Pathol* 1986; **1:** 3–14
17. Aretz HT: Myocarditis: the Dallas criteria. *Hum Pathol* 1987; **18:** 619–624