Natural History of the Idiopathic Cardiomyopathy

TSUTOMU INOH, M.D. AND ISAMU TAKESHITA, M.D.

Because of that the etiology of the idiopathic cardiomyopathy is still in obscure and the classification\(^1-3\) generally accepted has not been established pathologically\(^4\) and clinically, attempt to clarify the natural history\(^5\) is still in problem now. However, on the course of this disease sudden death of Stokes-Adams syndrome appears not uncommonly and the congestive heart failure is intractable once it has appeared, so that it is one of the urgent problem in clinical cardiology to figure out the natural history of this disease.

**Part 1: Idiopathic cardiomyopathy**

In this part natural history of the idiopathic cardiomyopathy has been investigated mainly on the autopnsed cases retrospectively.

Subjects: 51 autopsied and 55 living cases were subjected (Table I). In this part, familial cardiomyopathy, hypertrophic obstructive cardiomyopathy, endocardial fibroelastosis observed in infant and secondary cardiomyopathy were excluded. Autopsied cases were classified into two types as type I and II. Type II is with marked endocardial thickening and including the cases reported as endomyocardial sclerosis, endomyocardial fibrosis or adult type of endocardial fibroelastosis. Type I is without endocardial thickening and classified into three subdivisions such as Ia with prominent cell infiltration in the heart muscle, Ic with marked fibrosis and Ib as the intermediate type between Ia and Ic. Incidence of each type is as follows: 34 cases (66.7%) of type I, 11 cases (21.5%) of Ia, 9 cases (17.7%) of Ib, 14 cases (27.5%) of Ic and 11 cases (33.3%) of type II respectively. As the initial symptom palpitation and dyspnea were most frequent and fainting or syncope was also not uncommon. It is noticed that there is 6 cases

<table>
<thead>
<tr>
<th>Type of ICM</th>
<th>Autopsied</th>
<th>Living</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Ia</td>
<td>b</td>
</tr>
<tr>
<td>Male</td>
<td>5</td>
<td>7</td>
</tr>
<tr>
<td>Female</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>11</td>
<td>9</td>
</tr>
</tbody>
</table>

106 cases

**Key Words:** Cardiomyopathy  
Natural History  
Idiopathic Cardiomyopathy  
Familial Cardiomyopathy  
Endocardial Fibroelastosis

*Kobe University, School of Medicine, Department of Internal Medicine, Division I, Kobe, Japan*  
*This paper was presented as the Symposium 'Natural History of Congenital Heart Disease' on the 35th Annual Meeting of the Japanese Circulation Society in Tokyo, April 4, 1971.*

838 *Japanese Circulation Journal* Vol. 36, August 1972
died suddenly without any subjective complaint before their death. At the onset of the disease fever or signs suggesting the inflammation were observed in 27 cases of all subjects.

Averaged total course of the disease, which means duration from the onset of initial subjective symptoms to death, in each groups was 5 days in Ia, 21 months in Ib, 50 months in Ic and 60 months in type II respectively. Chronic congestive heart failure occurred most frequently in Ic and type II, while in Ia and Ib sudden death is more frequent than chronic congestive heart failure. Stokes-Adams attack was seen in any stage of the clinical course of all types and its incidence is 22% in total cases. Death occurred at any decade, however the death at the 4th decade was most frequent. Sudden death and congestive heart failure were major cause of death. Mean cardio-thoracic index (CTI) before death in each group was 0.51 in Ia, 0.65 in Ib, 0.63 in Ic and 0.68 in type II respectively. Enlarged hearts over 500g were seen in about a half of the cases with a long course of the disease over 18 months, however there were many cases died even with a small heart less than 400g of the heart weight especially in the group of early death and many of them belonged to Ia histologically. Majority of the type Ia died by acute course within 3 months.

Co-relations between the histological findings of the heart and its clinical course were analysed. Hypertrophy, degeneration and atrophy of muscle fibre were seen in every stage of the course of type I, and dominant cell infiltration was seen in the case of early death and marked fibrosis in the late death generally. However it was noticed that there were some cases with marked cell infiltration even with the course of more than one year. In the type II, marked thickening of the endocardium was observed invariably and cell infiltration was rather slight. The duration of the course is variable such as fluminent course died within 5 days from the onset and very chronic with fairly good prognosis (Fig.1). 27% of the 55 living cases were surviving over 10 years and 17% over 15 years and maximal course of our cases is 39 years from its onset. Calculated survival rate at 5th year from the sufficiently followed up patients to be calculated is 75% (Fig.2).

Summarized schema of the natural history of the idiopathic cardiomyopathy presented in Part I is shown in Fig.3. 25% of the cases began with fever or clinical signs suggesting acute inflammation such as positive CRP but the others were insidious. In this study calculated moratility rate was 17% including 2.4% with acute or fluminating course, 5.1% with subacute course and 9.5% with chronic course. As the cause of death sudden death is not uncommon especially in the group of early death or with acute course while the congestive heart failure is rather common in the chronic cases. According to the histological analysis hypertrophy, atrophy and degeneration of the heart muscle were common findings in each type of disease classified in this
Fig. 2. Survival rate calculated and duration of disease from its onset of the living subjects.

Fig. 3. Summarized schema of the natural history of the idiopathic cardiomyopathy (upper part) and the explanation of the histological findings of the heart on the course and types of the disease (lower part).

Japanese Circulation Journal Vol. 36, August 1972
study and at any stage of course, while remarkable cell infiltration was observed in Ia with acute course and generalized marked fibrosis in Ic with chronic course respectively.

Part II: Familial Cardiomyopathy

In this part 36 families of familial cardiomyopathy including 18 families from the Japanese literature were investigated. Among them 7 families of hypertrophic obstructive cardiomyopathy (HOCM) that is 19% of all families were included. 99 cases including 15 cases of familial hypertrophic obstructive cardiomyopathies were studied. 54 cases were male and 30 were female in non-obstructive familial cardiomyopathy. In familial hypertrophic obstructive cardiomyopathy 9 were male and 6 were female. In electrocardiographic findings of non-obstructive cardiomyopathy various arrhythmias were seen most frequently (in 21 cases) and other findings were as following that change of ST-T segment (in 16 cases), abnormal Q (in 16 cases), ventricular hypertrophy (in 13 cases) and paroxysmal tachycardia (in 8 cases) respectively. 26 cases showed cardiomegaly over 0.5 of car-
TABLE II  RELATION BETWEEN THE MAINLY MANIFESTED CLINICAL SIGNS AND CLINICAL COURSE OF FAMILIAR CARDIOMYOPATHY

<table>
<thead>
<tr>
<th></th>
<th>Sudden death</th>
<th>Living</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>CHF</td>
<td>Severe</td>
</tr>
<tr>
<td>Cardiomegaly</td>
<td>5</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Paroxy, Tachycard.</td>
<td>5</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>Block S-A A-V</td>
<td></td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Others</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>10</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>HOCM</td>
<td>1</td>
<td>1</td>
<td>5</td>
</tr>
</tbody>
</table>

Clinical course was classified into 4 groups such as sudden death, congestive heart failure (CHF) and in severe or mild clinical condition.

dio-thoracic index and 21 cases showed normal size of the heart. Onset of initial symptoms was one decade earlier than that of the non-familial cardiomyopathy reported in part I and most frequent at the 2nd decade. Onset of initial symptoms in familial hypertrophic obstructive cardiomyopathy was also most frequent at 2nd decade and this was as same as in non-familial hypertrophic obstructive cardiomyopathy. Pulpitation, arrhythmia and chest pain were the common symptom appeared at the onset of disease. It is noticed that the fairly many asymptomatic cases were found even in the family including severe cases. Prognosis and age of death is variable and different in each family. Sudden death was more frequent (75% of cardiac death) than that of the non-familial cardiomyopathy (40%) (Fig.4). Among the ten suddenly died cases of non-obstructive familial cardiomyopathy 5 cases revealed cardiomegaly in chest X-ray examination and in 5 cases the paroxysmal ventricular tachycardia in electrocardiogram examined before death was proven (Table 2). Of total 18 cases with cardiomegaly 5 cases died suddenly and 4 cases were in congestive heart failure or in other severe conditions, while 9, half of total cases with cardiomegaly, were in mild condition clinically.

Part III: Endocardial Fibroelastosis

Age of death of endocardial fibroelastosis and allied disease reported in Annual of the Pathological Autopsy Cases in Japan of recent 5 years were summerized in this part. Entity of the endocardial fibroelastosis seen in infant is established, however, there is still some confusion about the diagnosis of this disease seen in aged cases. The differentiation among the endocardial fibroelastosis seen in infant, adult type of it, endomyocardial sclerosis and endomyocardial fibrosis is in problem. In this report such group with endocardial thickening seen in aged cases was discussed in Part I. Of 55 cases died at first decade 21 cases were died within 6 months and 17 cases of them were accompanied with another heart anomalies, while in the cases survived over 6 months complication of another heart anomaly were rare. 18 cases were died within one year over 6 months, 9 cases within 5 years over one year and 7 cases within 10 years over 5 years respectively (Fig.5).

SUMMARY

Natural history of the idiopathic cardiomyopathy was studied. In part I idiopathic cardiomyopathy, excluding hypertrophic obstructive cardiomyopathy, familial cardiomyopathy, endocardial fibroelastosis seen in infant and secondary cardiomyopathy, was subjected. Subjects reported in Part I were classified into type I with 3 subdivisions and type II by histological findings of the heart and incidence and natural history of each type were discussed with comparing each other. Natural history is variable and different in each type such as fluminating and chronic. Calculated survival rate at 5th year was 75%.

In part II familial cardiomyopathy was subjected. In this group onset of initial symptoms

*Japanese Circulation Journal  Vol. 36, August 1972*
Fig. 5. Each column shows the number of cases of endocardial fibroelastosis and allied disease died until each age and solid line in the column means number of cases accompanied by another congenital heart anomalies.

was most frequent at 2nd decade that was earlier than the other non-familial group reported in part I. Age of death was variable in each case.

In part III endocardial fibroelastosis seen in infant obtained from the Annual of the Pathological Autopsy Cases in Japan was subjected. Majority of this group died within one year. 76% of the cases died within 6 months were accompanied by another heart anomalies but in the survived cases over 6 months or more complication of another heart anomaly was rather rare. Major causes of death through each type of cardiomyopathy reported in part I and II were sudden death and intractable chronic congestive heart failure. In familial cardiomyopathy sudden death had occurred at the rate of 75% that was much more frequent than the rate of 40% observed in the non-familial idiopathic cardiomyopathy reported in part I.

*Japanese Circulation Journal* Vol. 36, August 1972
Acknowledgement

The authors wish to express thanks sincerely to the Dept. of Medicine and the Dept. of Pathology in the Kinki area for their co-operation by presenting the valuable cases. The authors are grateful to the chairman of this symposium, Prof. Izumi and Associate Prof. Okada, and Prof. Tomonatsu for his advice.

The co-operation by Dr. M. Hatada, Dr. S. Takahashi, Dr. K. Kobayashi, Dr. T. Suzuki, Dr. K. Nakai, and Miss Namba is appreciated.

REFERENCES


