

한국에서의 소아기 원발성 심근증

천은정 · 강이석 · 배은정 · 이종균 · 길홍량 · 윤향석
김용욱 · 박희주 · 고재곤 · 노정일 · 이흥재

Idiopathic Cardiomyopathies in Korean Children : A Nationwide Study

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ABSTRACT

Background : Although idiopathic cardiomyopathies (i-CMP) are very important in all age groups, the epidemiology of i-CMP in children has not been well defined. A retrospective study in Korean children was performed in 1998 to obtain basic data on i-CMP. **Material and Method :** The medical records of all patients aged birth to 15 years from the hospitals where pediatric cardiologists worked were reviewed to obtain information on i-CMP. Pediatric cardiologists from a total of 22 hospitals were participated in reviewing the medical records of their patients and filling up the protocol. The data were pooled to the study committee and reviewed. **Results :** Of the 278 cases with i-CMP, there were dilated cardiomyopathy (d-CMP) in 182 (65.4%) ; hypertrophic cardiomyopathy (h-CMP) in 74 (26.6%) ; restrictive cardiomyopathy (r-CMP) and unclassified in 17 (6.1%) and 5 (1.9%) each. The average annual occurrence of new cases as a whole was 2.65 per 100,000 (95% CI ; 1.5 -3.7) ; d-CMP, 1.73/100,000/year (95% CI ; 0.73 -2.73) ; h-CMP, 0.71/100,000/year (95% CI ; 0.35 -1.07) ; r-CMP, 0.16/100,000/year (95% CI ; 0.02 -0.3). The median age at the time of diagnosis was 11 months in d-CMP ; 3.0 years in h-CMP ; 6.9 years in r-CMP. The survival rate in d-CMP was 76% at 1 year, 72.5% at 2 year, 70% at 5 year. There was no difference in survival rate according to age (in d-CMP, between children less than 2 years of age and over 2 years of age (74% vs. 79% at 1 year ; 67% vs. 76% at 5 year, p = NS) ; in h-CMP, between children less than 1 year of age and over 1 year of age (84% vs. 96% at 1 year ; 63% vs. 81% at 5 year, p = NS). R-CMP showed the worst survival rate (72% at 1 year, 30.2% at 5 year). **Conclusion :** In spite of the inherent defects of retrospective analysis, this study provides the useful epidemiological data in children with i-CMP. However, more systemic approach is needed to define the nature of the i-CMP in children. (**Korean Circulation J 2000;30(5):635-645**)

KEY WORDS : Idiopathic cardiomyopathy · Korean children · Epidemiology.

서 론

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가 . 가

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1980 (World Health Organization ; WHO) “

가
가

1)

대상 및 방법

1995

2)

0 15

(unclassified) (arrhythmic right ventricular cardiomyopathy)
가 가

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(multicenter

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study)

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1978 1987 10

46

10 (1988~1997) 278

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10 가

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9, 16, 40, 22, 11, 12

가

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10 10 (1988

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82% 22

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(Intel Pen -

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82% 22

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진단과 분류

2)

(Table 1).

Table 1. Classification of idiopathic cardiomyopathies (WHO 1995)

Dilated CMP
Hypertrophic CMP
Restrictive CMP
Arrhythmogenic Right Ventricular CMP
Unclassified
Fibroelastosis
Noncompacted myocardium
Systolic dysfunction with minimal dilation
Mitochondrial disease

21%

12%

278

242 (88%)

33 (12%)

3%

2 (stan - 가 . Doppler
dard deviation) ejec - 2/3
tion fraction 55 , fractional shortening 28 .⁹⁾
. fibroelastosis, noncompacted my -
ocardium, systolic dysfunction with minimal dilation,
mitochondrial disease가 . 1996 WHO
arrhythmogenic right ventricu -
lar cardiomyopathy
2 .
⁶⁾ , 가 ,
, anthracycline
, .
⁸⁾ .

Table 2. New cases of CMP in Korean children, 1988 - 1997

Type of CMP & age	No. of males	No./100,000	No. of female	No./100,000	Total No./100,000
DCMP					
0 - 5	57	3.13	81	4.97	4.05
5 - 10	12	0.56	9	0.54	0.59
10 - 15	10	0.37	9	0.47	0.51
Unknown	0		3		
Total	82	1.26	102	1.96	1.73
HCMP					
0 - 5	25	1.37	18	1.10	1.26
5 - 10	9	0.49	5	0.30	0.39
10 - 15	12	0.60	5	0.26	0.45
Total	46	0.80	28	0.54	0.71
RCMP					
0 - 5	5	0.27	2	0.12	0.21
5 - 10	4	0.27	4	0.24	0.25
10 - 15	1	0.05	0	0.00	0.03
Total	11	0.19	6	0.12	0.16
Unclassified					
0 - 5	2	0.12	3	0.16	0.14
5 - 10	0	0.00	0	0.00	0.00
10 - 15	0	0.00	0	0.00	0.00
Total	2	0.03	3	0.05	0.04
All CMP					
0 - 5	89	4.89	104	6.38	5.67
5 - 10	26	1.43	18	1.07	1.23
10 - 15	23	1.15	14	0.74	0.99
Unknown	0		3		
Total	138	2.41	139	2.67	2.65

자 료 분 석

SAS Package 6.1

P 0.05

ANOVA

1988~1997

T test

95

(Confidence Interval ; CI)

Poisson

10)

결 과

10 (1988~1997)

278

가

182 (65.4%)

74 (26.6%),

17 (6.1%), 5 (1.9%)

0~15 (

11),

0~15 (3.0

),

4 ~15 (6.9)

가 (p=0.003)

가

(Table 2).

15

10

2.65 (95% CI ; 1.5~3.7)

5

가

가

, 5

4.97(95% CI ;

3.8 ~6.0) : 3.13(95% CI ; 1.9~4.3)

(p<0.04).

(Figs. 1 and 2).

5

가

5

가

가

70%

가

가

1997 12 31

15

3,418,184

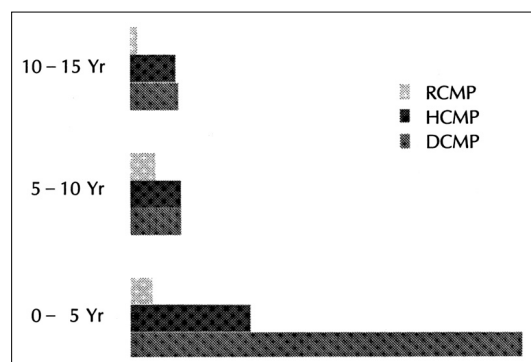


Fig. 1. Age distribution of CMP (I).

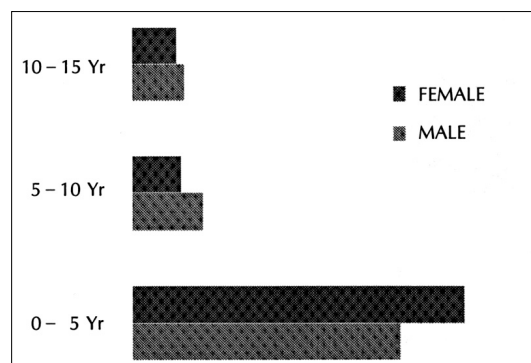


Fig. 2. Age distribution of CMP (II).

6.4~10.8) . 8.6 (95% CI ; 11)12)

확장성 심근증 182 (79, 102, 1)¹⁾ 1.73 /100,000 (95% CI ; 0.73~2.73) .

1.26 /100,000 1.96 /100,000 가 가 0~5 4.05 /100,000 가 (, 4.97/ 100,000 ; , 3.13/100,000) 0~15 11 가 가

90 가 (Fig. 3). 80 18 90 11 ~30 가 2.4 가

12 (Fig. 4). (skewness 0.08, $p=0.0001$).

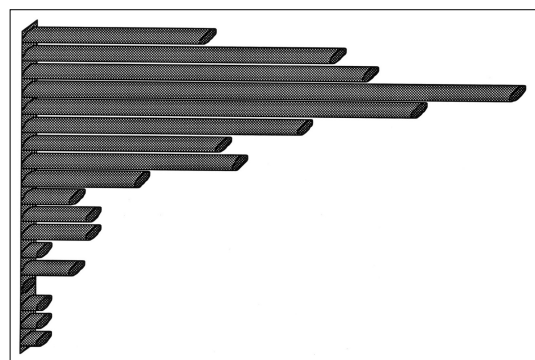


Fig. 3. Occurrence of new cases of i-CMP (I). IDC ; Idiopathic dilated cardiomyopathy.

1997 12 31 4.56 /100,000(95% CI ; 2.9~6.3) . 5 1988~1992 1993~1997 37 , 145 5 가 ($p=0.002$).

11 (7%) 102 (56%) , , (11%), (3%), (3%) 13 (7%) .

(4), (2), (2) 87 47% 9 (4.9%) 가 2.4

1 , 2 , 5 76%, 72.5%, 70% 가 가 2 1 , 5 74%, 67% 2 1 , 5 79%, 76% 가 ($p=NS$).

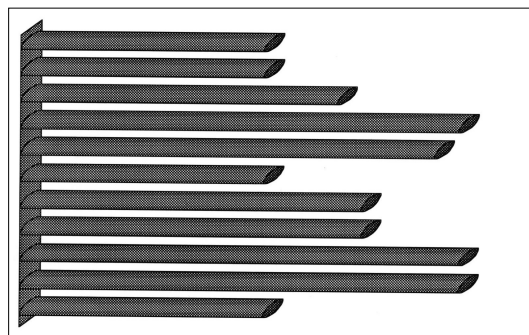


Fig. 4. Occurrence of new cases of i-CMP (II).

비후성 심근증

10 74 가
 0.71/100,000 (95%
 CI ; 0.35~1.07) 가 46 (62%) (76%) 가 56
 (28 , 38%)
 0~15 3.0 . 11 가 5 (6.7%)
 가 가
 1997 1.84 /100,000(95%
 CI ; 1.04~2.64)
 . 1988~1992 가
 32 (46%), 1993~1997 40 (54%) 5
 가 가
 가
 가 92%, 72%
 . 1
 1 , 5 84%, 63% 1
 96%, 81%
 . 가 16 (22%)
 14 (19%)
 4 (5%), 3 (4%)
 , 2
 가 29 (39%)
 20 (68%)
 가 5 (17%)
 3 가
 제한성 심근증
 10 (1988~1997)
 17 6%
 5 (1993~1997)
 가 14 (82%) 5 3 (18%)
 가 (p=0.03).
 가

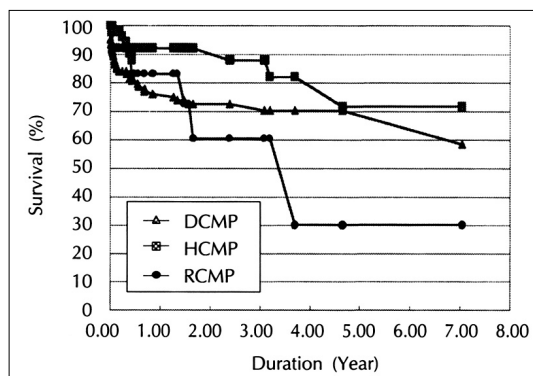


Fig. 5. Survival of children with cardiomyopathy.

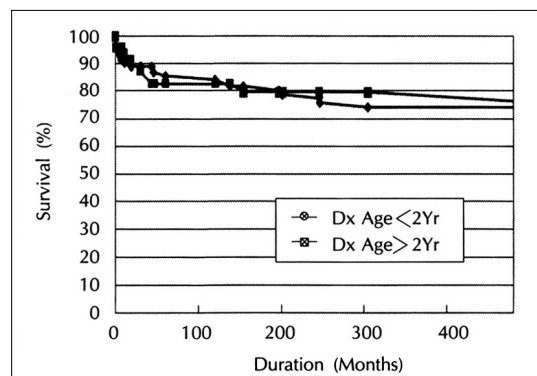


Fig. 6. Survival of children with dilated CMP.

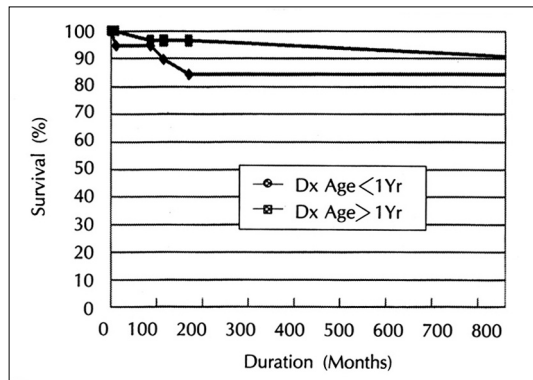


Fig. 7. Survival of children with hypertrophic CMP.

(multicenter study)

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가

M mode

가 10

가

0.16/100,000 (95% CI ; 0.02~0.3)

2, 3

10

(readily accessible)

(p = 0.04)

(13 76%). , 2 (11%),

1 (0.5%), 1 (0.5%)

3 8%

3 (18%)

가

6

(35%)

가

가

(p=0.02) 1 , 5

5 2/3

고 찰

가 가 /100,000(95% CI ; 0.73~2.73)

72%, 30.2%

가 Anita ¹⁶⁾ 100,000

0.34 Torpo ¹⁷⁾

10 10

가 . 6
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20)
2 가 2 (Fig.
0.71/100,000 (95%
CI ; 0.35~1.07) 0.17% Hada 6).
18)
Anita 10 0.34 가 (Fig. 7). 가
5 가 가 (p=0.02) 1 , 5
72%, 30.2% 5 2/3
1988~1992 32 (46%), 1993~1997
40 (54%)
가
가
가 가
가
22 2~3
17
5 (1993~1997) 가 14
(82%) 5 3 (18%) 가
(p=0.03). 21)22)
21%
가
가
가
173
39 1 , 5 76%,
70% 1 , 5
92%, 72% Henry 19)

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