

## Marfan

### Scoliosis Associated with Marfan Syndrome

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#### – Abstract –

**Objectives :** To evaluate characteristics and results of treatment of scoliosis associated with Marfan syndrome

**Methods :** The clinical data of 57 patients diagnosed as Marfan syndrome from January 1989 to June 1999 were reviewed. Thirty one patients had major curves more than 10 degrees by Cobb's method on whole spine standing film and their curve patterns and treatment methods were analyzed. Twelve cases of Marfan scoliosis who underwent posterior instrumented fusion were compared with randomly-sampled cases with idiopathic scoliosis about flexibility, correctability and correction loss of curves.

**Results :** Scoliosis was identified in 31 of 57 patients (54.4%) and among them 15 patients had the curve more than 40 degrees. Twelve curves were double major, 10 were thoracic, and 8 were double thoracic. Seventy-five per cent of the 57 patients had congenital heart disease and 40% had lens dislocation. Twelve of 31 cases underwent posterior fusion with pedicle screw instrumentation. Marfan scoliosis (mean flexibility; 39% in thoracic, 52% in lumbar) were significantly less flexible than idiopathic scoliosis (60% in thoracic, 93% in lumbar) ( $P<0.05$ ). Mean correctability of Marfan scoliosis of operation (61% in thoracic, 55% in lumbar) was poorer than that of idiopathic scoliosis (72% in thoracic, 67% in lumbar), however, this was not significant statistically ( $P>0.05$ ). Mean correction loss in Marfan syndrome was 3.2% at the mean 4.1 year follow-up in 9 cases.

**Conclusions :** The prevalence of scoliosis in Marfan syndrome shows positive correlation with severity of the disease. As compared to idiopathic scoliosis, the curve associated with Marfan syndrome was larger and less flexible. After posterior fusion using pedicle screw instrumentation, the correctability of Marfan curve was poorer than that of idiopathic curve, which is, however, insignificant statistically. So it was considered to be a good method to treat Marfan scoliosis.

**Key Words :** Marfan syndrome, Scoliosis

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1999

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(01-1999-019).

Marfan							
29 , 28							
22.6(5~58) . Marfan							
Pyertz McKusick <sup>10)</sup> Joseph <sup>5)</sup>							
. Marfan							
Marfan	,	,	,	,	(major sign;	,	,
,	,	,	,	,	,	(minor sign;	,
,	,	,	,	,	가	,	thumb sign,
,	,	가	,	,	wrist sign, knee joint sign)	1	(definite Mar-
,	,	,	,	,	fan syndrome; 2	가	),
. Marfan	,	,	,	,	2 (probable Marfan syndrome; 1		
가	,	,	,	,	)	3 (Marfan phenotype;	)
,	,	,	,	,	. Marfan		
가	가	가	가	가			,
4,7,14)		6)	Marfan				
Marfan							
Marfan							
Marfan							
12 (Table 1)							
가							
(							
16,17)							
12							
(Table 2).							
Marfan							
1 , 11 , 15.8(11~24)							
가 3							
9 , 14.8(11~21)							
Marfan							
1989 1 1999 6							
Marfan		57		가		. Marfan	
		. 57		2			

**Table 1.** Summary of operated cases of Scoliosis Associated with Marfan Syndrome

Case	Sex/Age	Curve* Pattern	Level	Fusion	Pre-Op.(°)	Side- Bending(°)	Post-Op.(°)
1	M/15	ST	T5-T12	T4-L1	66	30	7
2	F/15	ST	T5-T10	T4-T11	52	35	11
3	F/14	DT	T1-T7-L1	T2-L2	45/44	32/21	16/12
4	F/17	DT	T1-T6-T11	T1-T11	55/49	40/35	28/24
5	F/13	DT	T1-T7-T12	T1-L2	40/33	21/2	4/3
6	F/11	DM1	T6-T10-L3	T10-L2	35/66	20/58	27/43
7	F/13	DM1	T5-T11-L4	T5-L5	51/61	21/20	10/10
8†	F/17	DM2	T4-T12-L4	T4=L4	83/60	68/27	33/28
9‡	F/24	DM2	T6-L1-L5	T6-L3	96/53	68/9	21/10
10‡	F/12	DM2	T6-T12-L4	T5-L4	91/83	65/39	55/46
11‡	F/23	DM2	T5-T11-L4	T4-L4	78/68	54/27	29/28
12	F/16	DM2	T5-T12-L4	T4-T12	98/62	80/21	53/46

\* ST = single thoracic; DT = double thoracic; DM1 = double major (King 1);

DM2 = double major (King 2).

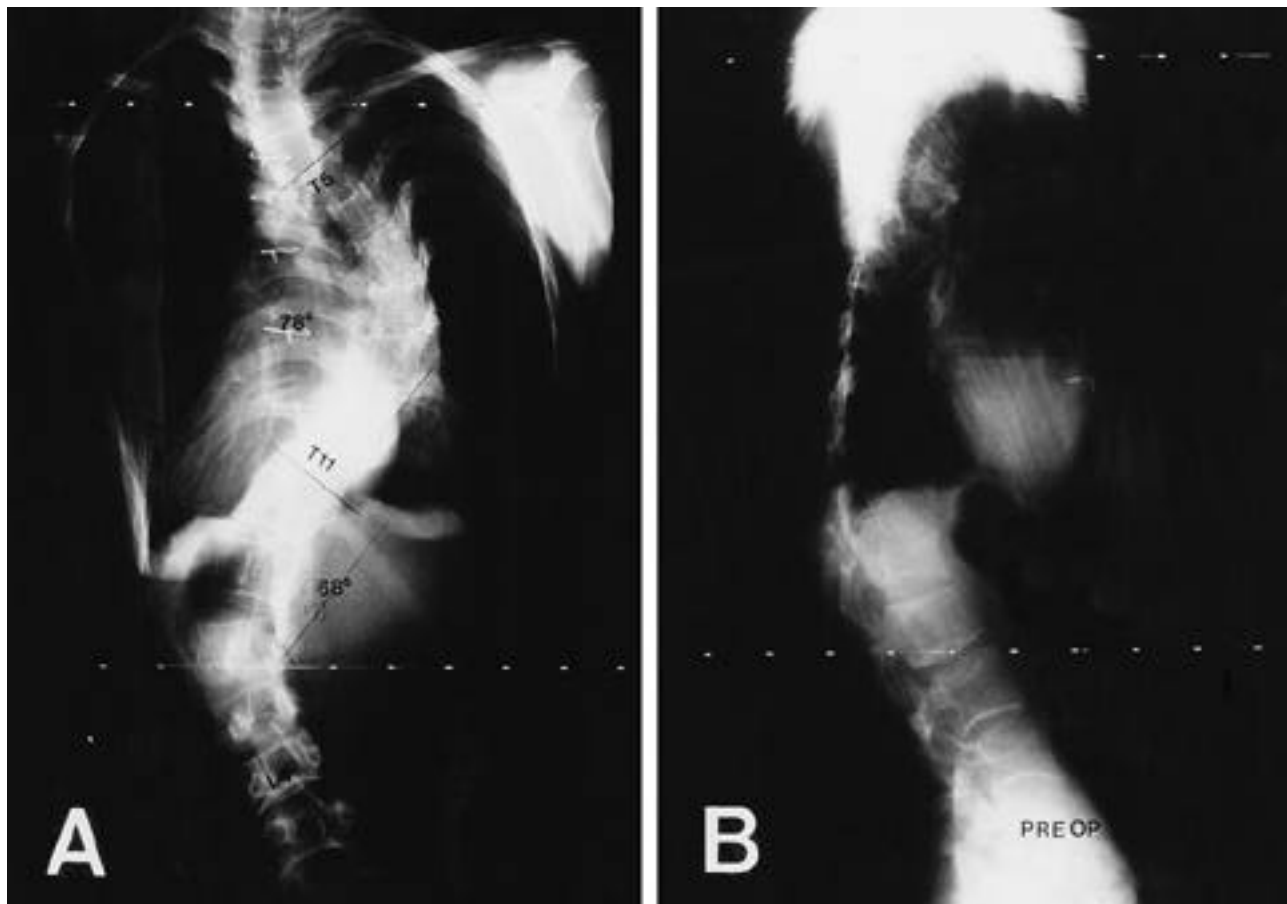
† Patients done anterior release and thoracoplasty.

‡ Heparinization

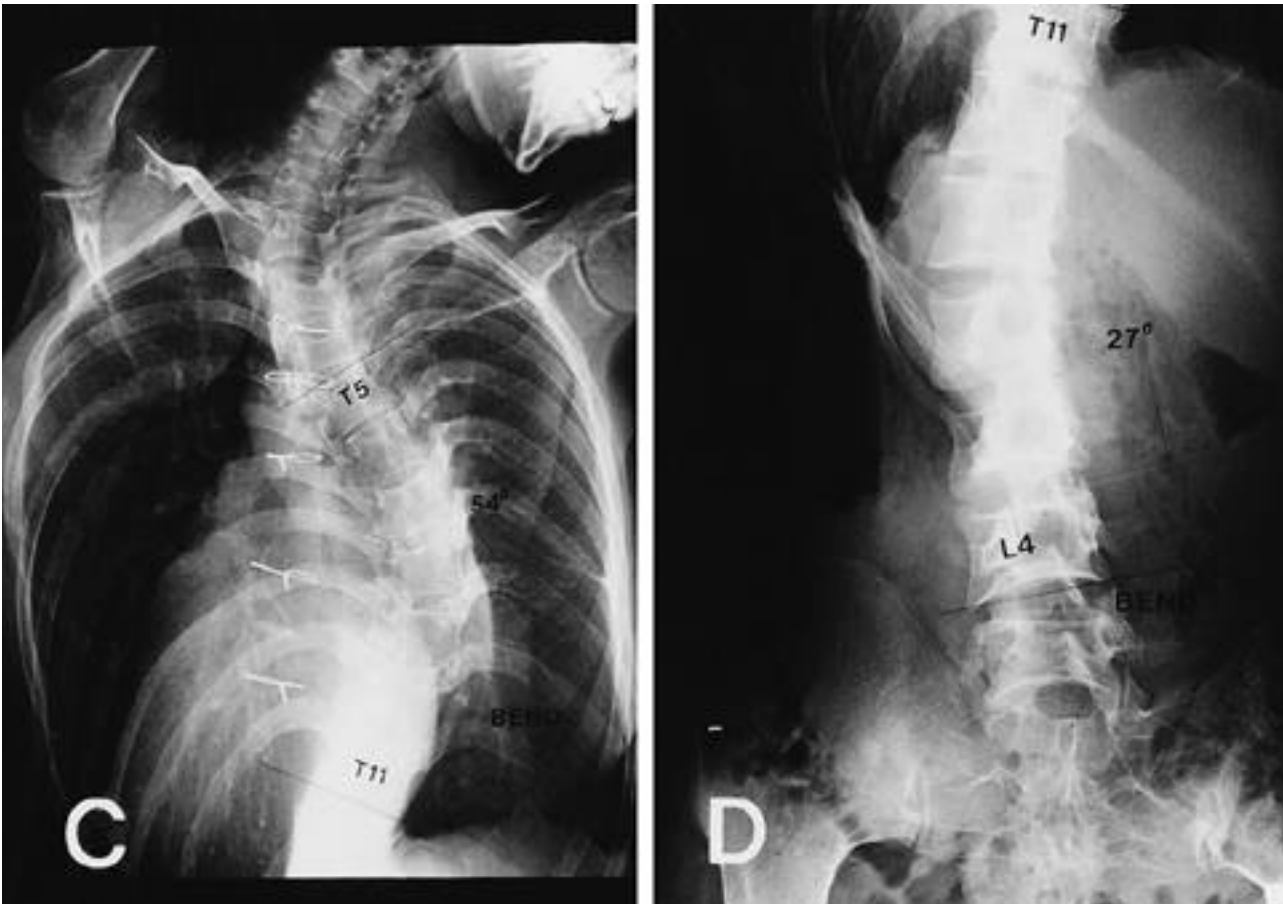
**Table 2.** Summary of operated cases of Idiopathic Scoliosis (sample)

Case	Sex/Age	Curve Pattern	Level	Fusion	Pre-Op.(°)	Side-Bending(°)	Post-Op.(°)
1	F/15	ST	T6-L1	T5-T12	41	14	5
2	M/15	ST	T5-T12	T5-T12	59	24	24
3	F/15	DT	T2-L3	T1-T6-L2	47/55	35/38	17/18
4	F/13	DT	T2-T12	T1-T6-T12	23/45	10/15	5/6
5	M/20	DT	T2-L3	T1-T6-L2	35/49	13/0	16/12
6	F/11	DM1	T5-L3	T7-T12-L3	45/61	15/19	18/10
7	F/13	DM1	T4-L3	T4-T10-L4	34/61	15/24	10/17
8	F/15	DM2	T4-L3	T4-T11-L3	63/62	26/18	12/20
9	F/11	DM2	T5-L3	T5-T11-L4	49/47	20/7	14/8
10	F/21	DM2	T6-L3	T6-L1-L5	44/22	8/-15	12/8
11	F/12	DM2	T4-T11	T5-T12-L4	51/41	13/-6	11/26
12	M/16	DM2	T4-T12	T6-T11-L4	67/31	45/5	15/10

\* ST = single thoracic; DT = double thoracic; DM1 = double major (King 1)  
DM2 = double major (King 2).



**Fig. 1-A,B.** A 22-year old girl with scoliosis associated with Marfan syndrome. She underwent an operation for heart disease and had been taking Coumadin(anticoagulant). The magnitudes of right thoracic and left lumbar curves were 78° and 68° retro-spectively.



**Fig. 2-C,D.** Side-bending radiographs. The thoracic and lumbar curves were corrected to 54° (31%) and 27° (60%).

hump)가 , (rib  
Cobb ,  
(Fig. 1A, B),  
(Fig. 2C, D),  
(Fig. 3E, F)  
Marfan  
Mann-Whitney  
test ,  
1 가 9  
Marfan  
(75%, 43/57) 가  
(40%, 23/57),  
Marfan 57 1 (definite Marfan  
syndrome) 24 , 2 (probable Marfan syndrome)  
28 , 3 (Marfan phenotype) 5 . 10  
31 ( 12 , 19 )  
54.4% ,  
definite Marfan syndrome 1 70.1%(17/24)  
2 (50.0%, 14/28), 3 (40.0%, 2/5)  
가 40 가 1  
37.5%(9/24), 2 21.4%(6/28) , 3  
(75%, 43/57) 가  
(40%, 23/57),

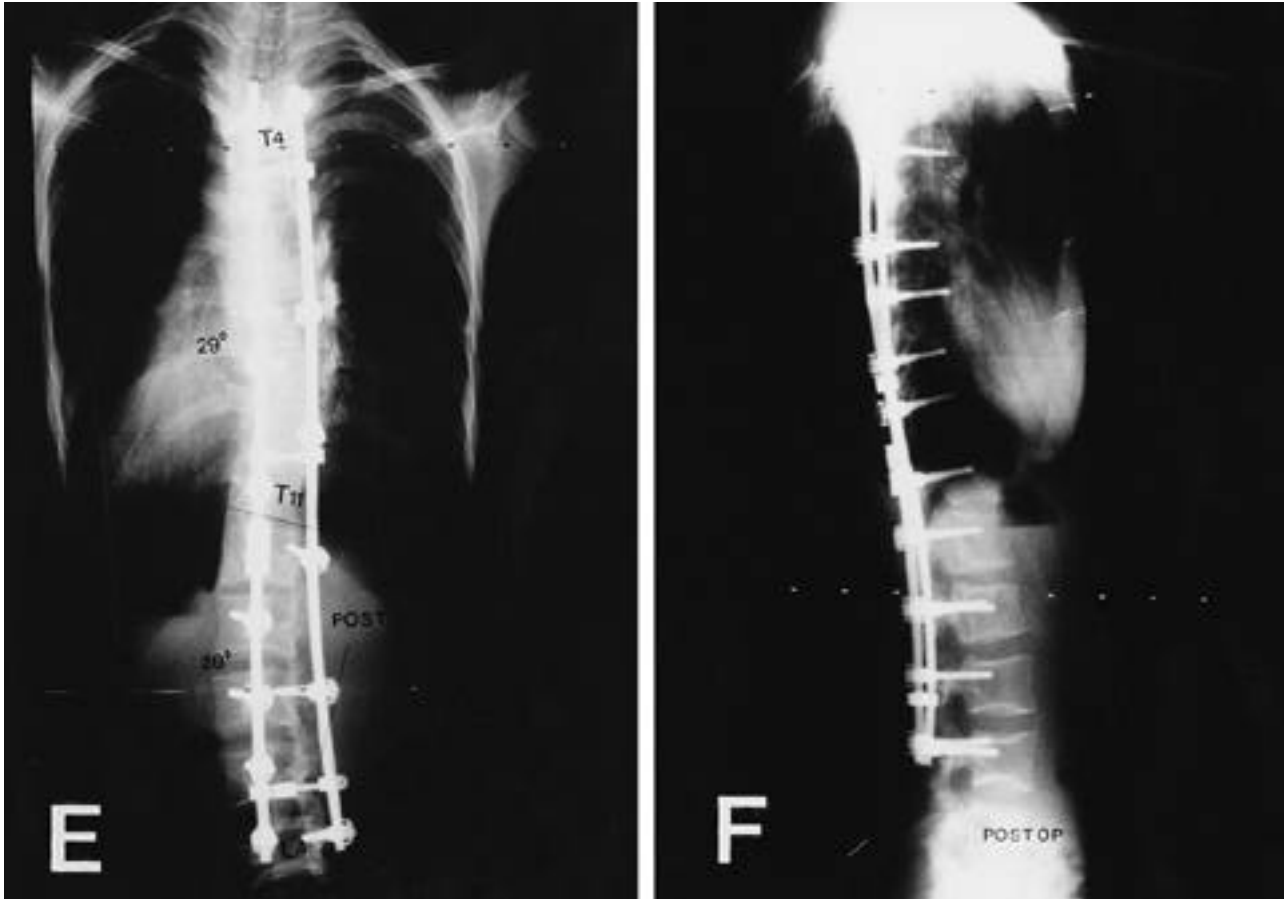


Fig. 3-E,F. Postoperative standing radiographs. The thoracic and lumbar curves were corrected to 29° (63%) and 28° (59%).

Table 3. Accompanied diseases of Marfan Scoliosis

Disease	No. of patient (%)
Heart disease	43 (75)
Lens dislocation	23 (40)
Thoracic hypokyphosis	8 (14)
Thoracic deformity	5 (8)
Spontaneous pneumothorax	2 (3.5)
Inguinal hernia	2 (3.5)

Table 4. Curve pattern of Marfan Scoliosis

Curve pattern	No. of patient
Double major	12
Single thoracic (Lt. : Rt.)	10 (2 : 8)
Double thoracic	8
Single lumbar	1

(14%, 8/57), (5%), (2%), (2%)  
(Table 3). 12 (100%)  
, 10  
( : =2:8), 8, 1  
(Table 4), 10, 20, 가 11,  
20, 40, 가 8, 40, 15,  
16, 3  
가 19, 가 12,  
3, 2  
가, 1  
.

5.  
Marfan  
15.  
fan  
~50° (21.6°) 39.4 ± 19.5%,  
8°~44° (29.9°) 51.8 ± 22.9%  
,  
12°~38° (26.6°) 59.9 ± 19.2%,  
26°~49° (36.9°) 93.1 ±  
37.3% Marfan  
(P<0.05). Marfan

Marfan									
8 <sub>o</sub> ~75 <sub>o</sub> ( 40.8 <sub>o</sub> )	61.4±	가	Marfan	42	29 (69%)				
23.1%, 9 <sub>o</sub> ~51 <sub>o</sub> ( 30.6 <sub>o</sub> )					, Robins <sup>11)</sup>	45			
54.7±21.8%			27		60%				
18 <sub>o</sub> ~52 <sub>o</sub> ( 33.2 <sub>o</sub> )					, Orcutt DeWald <sup>9)</sup>	73%			
72.3±10.0%, 14 <sub>o</sub> ~51 <sub>o</sub> ( 33.9 <sub>o</sub> )					Marfan				
67.5±16.1%	Marfan		57	10					
가			가 31						
(P>0.05).			54%						
Marfan	57			1	73%,	2			
가 30 (53%)	, Marfan		50%,	3	40%	, 40			
12				1	2	Marfan			
가 2									
					, Marfan				
2	10			가 29:28					
(level)	35.8	, 35.0		가	12:19,				
( 24 )					1:11				
0.89 pints, 0.82 pints			가						
	Marfan								
					가 가				
				<sup>19)</sup> . Tanesa Manning <sup>18)</sup>					
, 1			가		, Robins <sup>11)</sup>				
	12		48%가		가				
6	8 1	( 4.1 )	가	33%가					
9		3.2%(0~10.6%,	. LeDellieu <sup>19)</sup>	Sponseller <sup>12)</sup>	Marfan				
2.2 <sub>o</sub> (0~7 9)					가				
				, Winter <sup>21)</sup>	Mankin <sup>8)</sup>				
			가						
Marfan	0.01%	가	31						
3/4		1/4	가						
	<sup>19)</sup> ,				10				
15	long arm	fibrillin gene			2			8	
	<sup>3)</sup> ,								
Joseph <sup>5)</sup>	36	Marfan	<sup>15)</sup>			5			
1 (definite Marfan syndrome)	가 18		가			4.68%			
(50%), 2 (probable Marfan syndrome)	9 (25%),		10		2.28%, 20	0.27%, 30			
3 (Marfan phenotype)	9 (25%)		0.08%						
	1 24	42%, 2 28			20				
49%, 3 5	, 9%	Joseph				Marfan			
3					40	48%, 10			
			20	32%					
Marfan	40~60%		가	가		Marfan			
<sup>4)</sup> . Markin <sup>8)</sup>	2								

Marfan . Marfan  
 가 <sup>19)</sup>. Birch Herring<sup>1)</sup> Boston, Milwaukee , 50%  
 가 9 8 80% 가  
 , Robins <sup>11)</sup>  
 Marfan (mitral valve prolapse) 가  
 Milwaukee 가 <sup>19)</sup>  
 (rigid) , 가 가 43 75%  
 40%, 9%  
 Milwaukee TLSO 가 가  
 가  
 8  
 가 2 6  
 5 1 (heparinization)  
 가 ( 24 ) ,  
 가  
 Brenton <sup>2)</sup> Marfan  
 , elastin  
 , Winter <sup>20)</sup> Marfan  
 . Marfan  
 (ligamentous  
 laxity) 가  
 (rigid) <sup>19)</sup>  
 Sponseller <sup>12,13)</sup> Marfan  
 (pathogenetic mechanism)  
 Marfan  
 12 Marfan  
 Marfan 가  
 fan <sup>4,11,19)</sup> Mar-  
 17)  
 가 58 ° 48%  
 72%, 30 71%  
 12 Marfan  
 Marfan

가

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54.4% , Marfan 가

가 가 ,  
가 가 ,

Marfan

가 ,

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: Marfan , 가 ,

: Marfan , , ,

: 1989 1 1999 6 Marfan 57 Cobb , 10

31 12 , 12

: Marfan 54% , 가 40

48% . (75%), (40%) ,

12 , 10 , 8 31 12

. Marfan 39%, 52% 60%,

93% (P<0.05). Marfan 61%,

55% 72%, 67% (P>0.05). 6

3.2% .

: Marfan 가 가 가 .

: Marfan ,

: