

## 수정 대혈관 전위 환아의 중기추적 관찰

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## Midterm Follow-up of Children with Corrected Transposition of the Great Arteries

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## ABSTRACT

**Background and Objectives :** Corrected transposition of the great arteries (C-TGA) is a rare congenital heart disease, of which prognosis depends on the associated cardiac defects, systemic ventricular function, competency of atrioventricular valves, and the presence of conduction disturbances. This study was aimed to assess the midterm follow-up status of C-TGA. **Patients and Methods :** Retrospective review was performed on 89 cases with C-TGA and two ventricles of adequate size, which were diagnosed between January 1980 and June 1997. **Results :** Study subjects consisted of 56 males and 33 females (average age at diagnosis, 9 months). Mean follow-up duration was 98 months (range, 2 months-23 years 8 months). Based on the associated cardiac anomalies, there were 6 simple C-TGA and 83 complex C-TGA patients. Surgery including 19 palliative and 47 corrective operations was attempted on 61 cases at mean age of 69 months. Tricuspid regurgitation (TR) was noted at the time of first examination in 52 (mild in 39 ; moderate in 8 ; severe in 5) and progressed in 18 patients. TVR was done on 5 patients and double switch on 7 patients. Arrhythmia was noted preoperatively (complete AV block in 3) in 11 and postoperatively (postoperative complete AV block in 3) in 22 patients. A total of 13 cases died including 10 perioperative deaths during follow-up. Actuarial survival rate at 10 year was 84.5%. **Conclusion :** In this study, the midterm outcome of corrected TGA is acceptable. However, long-term follow-up is required in respect to the function of atrioventricular valve and the systemic ventricle. (**Korean Circulation J 1998;28(10):1774-1781**)

**KEY WORD :** Corrected transposition of the great arteries.

## 서 론

eat arteries ; C - TGA) Von Rokitsky

1%

(corrected transposition of the gr -

.<sup>1)</sup>

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28

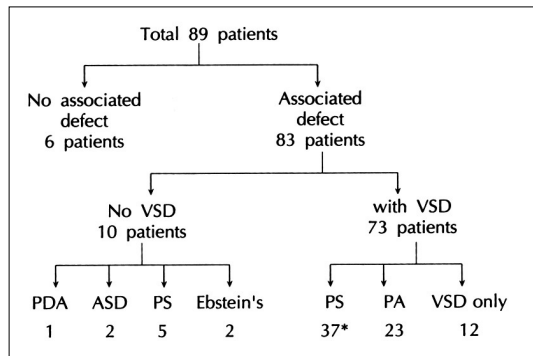
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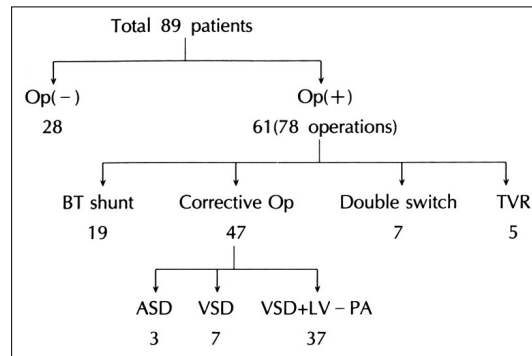
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double switch  
가  
가  
double switch  
가  
2)  
24 Holter  
24 Holter  
(C - TGA)  
SAS program Kaplan - Meier  
대상 및 방법  
1980 1 1997 6  
89 임상 양상  
89 56 33  
가 38  
가 6  
가 45 9 ( 1 ) 0 8  
가 8 2 ( 2 23  
가 8 )  
심기형의 동반 양상(Fig. 1)  
3  
가 color Doppler pulsed Do -  
ppler 가 1/3 C - TGA가 6  
가 1/2 가 C - TGA가 83  
가 1/2 가  
가 solitus, D -  
loop, L - position {S,L,L}  
가 74 , 가 inversus, D - loop,  
D - position {I,D,D} 가 15  
(ejection fraction)  
(contractility)  
가 73  
38 , 가 가 23  
가 8 2 ,  
1 , 5



**Fig. 1.** Associated cardiovascular anomalies in 89 children with corrected transposition of great arteries. ASD, atrial septal defect ; PDA, patent ductus arteriosus ; VSD, ventricular septal defect ; PS, pulmonary stenosis ; PA, pulmonary atresia ; \*, include 4 patients with Ebstein's anomaly.

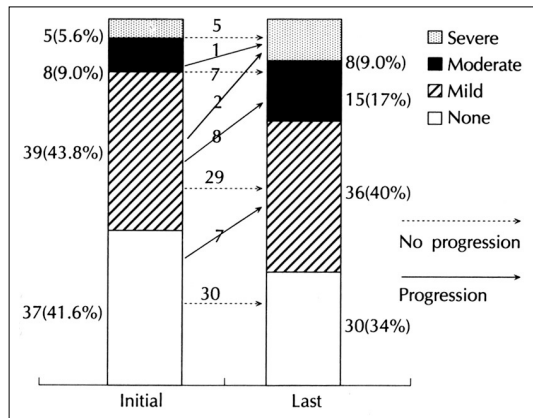


**Fig. 2.** Surgical treatment for corrected TGA patients. ASD, atrial septal defect ; PDA, patent ductus arteriosus ; VSD, ventricular septal defect ; PS, pulmonary stenosis ; PA, pulmonary atresia ; LV-PA, left ventricle to pulmonary artery ; TVR, tricuspid valve replacement ; Op(-), no operation ; Op(+), operation performed ; pts, patients.

Ebstein 6 , 113 , 82 )  
 가 2 ,  
 가 4 .

수 술 (Fig. 2)  
 61 .  
 5 9 , 2 14 4  
 .  
 (Blalock - Ta -  
 ussig shunt) 19 47  
 . 19 16  
 .  
 가 3 ,  
 7 ,  
 가 37 .  
 7 double switch 5  
 , 2 .  
 , 19  
 , 10  
 5 C - TGA  
 가 2 , C - TGA가 3 1 ,  
 2 . 4  
 , 1  
 7 .  
 27, 98, 98, 128, 130 . Ebs -  
 tein 6 ( 25

삼첨판 역류  
 가 가 52  
 , 가 39, 8, 5 .  
 13 C -  
 TGA가 4 , C - TGA가 9  
 1 ,  
 가 3 , 가 2 , 2  
 . Ebstein 3 2  
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 89  
 18 .  
 37 7  
 가 .  
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 , 가  
 8 , 가 2 ,  
 가 1 (Fig. 3).  
 41 160 ,  
 20 146 ,  
 27 42 .  
 5 ,



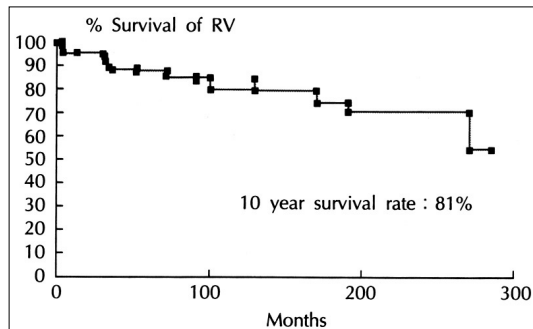
**Fig. 3.** Initial and follow-up status of tricuspid regurgitation were depicted showing the progression of TR in 18 patients.

**Table 1.** Arrhythmia in children with corrected transposition of the great arteries

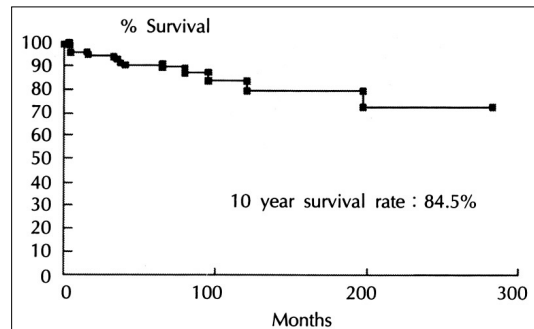
Preoperative (n=11)		Postoperative (n=22)	
Sinus pause	1	Sinus node dysfunction	1
Atrial flutter	1	Paroxysmal atrial tachycardia	1
WPW syndrome	1	Premature atrial contraction	5
Atrioventricular block			
1 <sup>st</sup> degree	4		
3 <sup>rd</sup> degree	3	Atrioventricular block	
		1 <sup>st</sup> degree	2
		2 <sup>nd</sup> degree	4
		3 <sup>rd</sup> degree	3
Ventricular tachycardia (nonsustained)	1	Premature ventricular contraction	6

부정맥 (Table 1)

11, 2, 10  
1, 1, 1, 4, 10  
3, WPW syndrome, C-TGA, 1  
가 1, 가 1  
3  
C-TGA가 1, 1  
22, 5, 25, 가 17, 가 47  
1, 1, 2, 2, 1, 4, 17  
3, 6, 가 9, 가, 가 7, 가 1  
patch closure, 1, 10  
patch closure, patch closure, 가 4, 16  
1, patch closure, conduit, double switch, 5, patch, 8, 1  
1, patch, 가, 10, 81%  
type 4, VVI type 2, DDD, (Fig. 4), 76  
3, 2, 4, NYHA class, 17, 13  
1, 3, 10, 4



**Fig. 4.** Functional survival curve of the right ventricle in corrected transposition of the great arteries.



**Fig. 5.** Actuarial survival curve of the patients with corrected transposition of the great arteries.

**Table 2.** Characteristics of 13 mortality cases

Case	Sex	Associated defects	Age at Op	Operation	Age at death	Cause of death
1	F	VSD, PS		ND	3 mo	Ventricular failure
2	M	VSD, PS	2 mo	Double switch	2 mo	Ventricular failure
3	F	VSD, PA	2 mo	VSD patch closure, LV-PA connection	2 mo	Ventricular failure
4	M	VSD, PA	4 mo	Double switch	4 mo	Ventricular failure
5	M	PS		ND	1 yr 10 mo	ARF, Reye syndrome
6	F	VSD, PS	2 yr 1 mo	VSD patch closure, LV-PA connection	2 yr 2 mo	Ventricular failure
7	F	VSD, PA	1 yr 7 mo	VSD patch closure, LV-PA connection	2 yr 3 mo	Ventricular failure
8	F	VSD, PS	2 yr 8 mo	VSD patch closure, LV-PA connection	2 yr 8 mo	Ventricular failure d/t coronary artery injury
9	M	VSD, PS	4 yr 10 mo	VSD patch closure, LV-PA connection	4 yr 10 mo	Ventricular failure
10	F	VSD, PS	6 yr 3 mo	VSD patch closure, LV-PA connection	6 yr 4 mo	Ventricular failure
11	M	VSD, PA	7 yr 7 mo	Double switch	7 yr 7 mo	Ventricular failure
12	M	VSD, PA	10 yr 5 mo	VSD patch closure, LV-PA connection	10 yr 5 mo	Ventricular failure
13	M	VSD, PA	16 yr	VSD patch closure, LV-PA connection	16 yr 1 mo	Ventricular failure

VSD, ventricular septal defect ; PS, pulmonary stenosis ; PA, pulmonary atresia ; LV-PA, left ventricle to pulmonary artery ; yr, year ; mo, month ; M, male ; F, female ; ND, not done ; d/t, due to

(NYHA class )

, 2

경 과

10

84.5%

13 . 10

(Fig. 5).

3

Double switch

7

,

8

5

3

2

2

Reye

16, 19, 67, 75

(Table 2). 13

C - TGA

(NYHA Class

C - TGA 6

).

고 안 , 5 가 , 17 가 , 4 . 가 , 3) . 4) switch Senning double - Mustard , Rastelli 가 가 . 가 , . 9-11) 7 double switch (ejection fraction) 가 4 가 . 가 가 가 5) 가가 . 6) , 10% 가 가 Fyler<sup>12)</sup> 20 가 45% 가 2% . 13) Elongated AV bundle 가 14) cooling cardioplegia . Connelley 5) 58 9 , 9 15 (36%) 7) 가 가 가 8) 가 , 가 가 . 가 7) 가 23 5 Huhta 7)

5 70%, 10 64% , 가 47 ,  
 . Lundstrom 25 , 가 17 13  
 15) 20 111 10 10  
 26 84.5%  
 McGrath 16) 99 1 결 론 :  
 75%, 10 68%  
 52  
 25% ,  
 가 5) 가 13  
 , 10 84.5%  
 ( 8 2 )

중심 단어 :

(98 - 026)

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## 요 약

연구배경 :

(C - TGA)

방 법 :

C - TGA 89

결 과 :

56, 33 9  
 8 2 C -  
 TGA가 6 , C - TGA가 83  
 61 5 9 ( 2  
 14 4 ) 19 ,  
 47 (TR)가  
 39, 8, 5 , 18  
 TR 5 , dou -  
 ble switch가 7  
 11 ( 3), 22 ( 3)

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