—**Original**—

Surgical Treatment of Coarctation Complex in Neonates and Infants

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Abstract

Background: There remains controversy regarding the appropriate surgical treatment of coarctation of the aorta associated with intracardiac anomalies in neonates and infants. Furthermore, the relative benefits of one versus two-stage repair, and subclavian flap aortoplasty versus end-to-end anastomosis for some of these lesions, remain controversial. The purpose of this paper is to review our experience with two-stage repair using subclavian flap aortoplasty and to seek an appropriate procedure.

Methods and Result: From June 1996 to November 1999, thirteen patients underwent subclavian flap aortoplasty in our department. The age range was 16 to 101 days (mean 52 days), and the body weight range was 1.9 to 4.5 kg (mean 3.0 kg). Anatomic diagnosis was coarctation with ventricular septal defect (six patients), double outlet right ventricle (two patients), atrioventricular canal defect (one patient), tricuspid atresia (two patients), mitral atresia (one patient), and single atrium and subaortic stenosis (one patient). There was one hospital death in our series due to the progression of pulmonary hypertension 3 months after the operation. The mean follow up for remaining twelve patients was 28 months (range 7～48 months). There was one reoperation for recurrent coarctation. Three patients underwent pulmonary artery plasty in a second operation because of right pulmonary artery stenosis. We performed the definitive operation for six patients with coarctation with ventricular septal defect and two patients with double outlet right ventricle, and we performed a bidirectional cavopulmonary shunt for four univentricular hearts who are candidates for the Fontan operation. Two patients required Damus-Kaye-Stansel procedure to release restrictive bulboventricular foramen. Three patients underwent a modified Fontan operation after these palliations. In our series, the intraoperative mortality rate for subclavian flap aortoplasty was 0% and the post operative mortality rate was 7.7% (1/13). Ten patients underwent the final operation successfully, and further two patients are considered good candidates for the final operation. The overall mortality was 7.7% (1/13).

Conclusion: Two-stage repair appears to offer a good prognosis for neonates and infants with a coarctation complex. Subclavian flap aortoplasty showed the lowest rate of restenosis. However, late mortality may be associated with the progression of pulmonary vascular disease and the presence of associated severe cardiac anomalies. Although Fontan candidates need staged operations, if biventricular repair is feasible, one-stage repair would be a reasonable procedure considering the progression of the pulmonary vascular disease and the distortion of the pulmonary artery due to pulmonary artery banding. It would appear to improve the quality of life of those children if a one-stage operation can be performed with reasonable risk and good midterm outcome. (J Nippon Med Sch 2000; 67: 455—458)

Key words: coarctation complex, two-stage repair, subclavian flap aortoplasty

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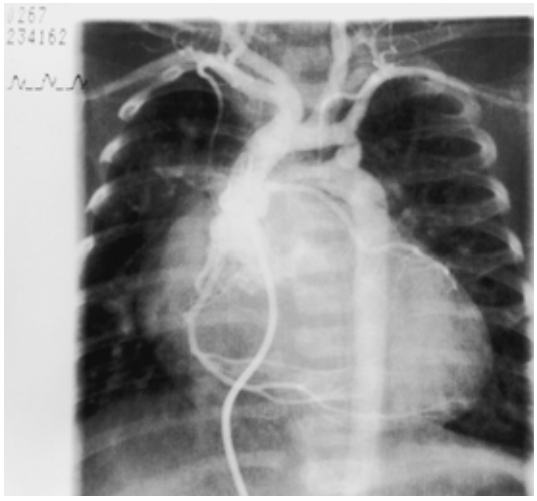

Fig. 1 Preoperative angiography of case 9 diagnosed as DORV, Taussig-Bing anomary, mitral stenosis, Coarctation of aorta and hypoplastic aortic arch.

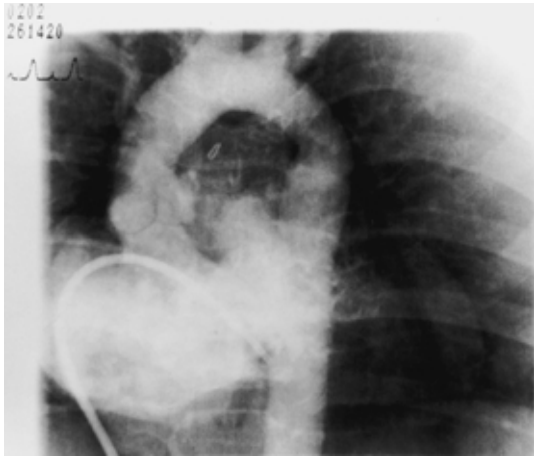

Fig. 2 Postoperative angiography of case 9 on whom subclavian flap aortoplasy was performed.

Introduction

The quest for an optimal surgical procedure for coarctation of the aorta began with Crafood and Nylin[1] in 1945, when they repaired an aortic coarctation in an infant by end-to-end anastomosis. More than 50 years later, controversy still exists as to the ideal management of this problem, especially in neonates and infants. Kirklin and associates[2] reported the successful repair of an aortic coarctation in the case of an infant with severe congestive heart failure using a similar technique. But the high recurrence rates with resection and end-to-end anastomosis prompted the development of the subclavian flap angioplasty technique for the surgical repair of infant coarctation first introduced in 1966 by Waldhausen and Nahrwold[3]. Advocates of this procedure claim that the absence of a posterior suture line aids in preventing the constriction and retraction associated with a circumferential anastomosis. Additionally, the subclavian flap is well vascularized and therefore has good growth potential (**Figs. 1. 2**). But the left thoracotomy for repair of the arch is complicated by the poor exposure of the proximal arch and a marked difficulty in palliating more complicated lesions such as transposition of great arteries and truncus arteriosus. Furthermore, for patients with a complex coarctation of the aorta plus a hypoplastic transverse arch, both the operative mortality and the recoarctation rate are higher if the hypoplastic segment is disreguarded, and residual obstruction will compound left ventricular dysfunction. This situation has stimulated interest in the one-stage approach now advocated for interrupted aortic arch in most centers. The best approach for coarctation of the aorta, however, remains controversial.

Materials and Methods

Patients: From June 1996 to November 1999, thirteen patients underwent subclavian flap aortoplasty. The age range was 16 to 101 days (mean 52 days), and the body weight range was 1.9 to 4.5 kg (mean 3.0 kg). The anatomic diagnosis was coarctation with ventricular septal defect (six patients), double outlet right ventricle (two patients), atrioventricular canal defect (one patient), tricuspid atresia (two patients), mitral atresia (one patient) and single atrium and subaortic stenosis (one patient). The patients' characteristics are summarized in **Table 1**. There was one hospital death in our series due to the progression of pulmonary hypertension 3 months after the operation in patient who was diagnosed as having single atrium, subaortic stenosis and congenital atrioventricular block and underwent subclavian flap aortoplasty, patent ductus arteriosus ligation and pacemaker implantation without pulmonary artery banding. The mean follow up for another twelve patients was 28 months (range 7～48 months). There was one reoperation for recurrent coarctation and one patient needed balloon

Table 1 Patient data

case	sex	age(day)	Bw(Kg)	diag.	follow(m)	additonal procedure	final operation
1	m	101	4.0	VSD	48	re CoA repair	ICR
2	f	70	2.5	VSD	44	(−)	ICR
3	m	43	2.9	VSD	38	PA plasty	ICR
4	m	41	3.0	TA(IIc)	37	BCPS+DKS	TCPC
5	f	32	2.9	MA	31	BCPS+PA plasty	TCPC + DKS
6	f	16	3.0	DORV	29	(−)	ICR
7	f	20	2.4	TA(Ic)	25	BCPS+PA plasty	awaiting TCPC
8	f	24	1.9	VSD	23	(−)	ICR
9	f	61	3.3	DORV MS	22	BCPS	TCPC
10	f	81	2.9	SA LVOTO AVB	dead	(−)	(−)
11	m	80	4.5	VSD	18	(−)	ICR
12	m	59	3.5	VSD	13	Balloon angioplasty	ICR
13	f	49	2.7	AVC	7	(−)	awaiting ICR
mean		52	3.0		28		

VSD: ventricular septal defect, TA: tricuspid atresia, MA: mitral atresia, DORV: double outlet right ventricle, MS: mitral stenosis, SA: single atrium, LVOTO: left ventricular outflow tract obstruction, AVB: atrioventricular, block, AVC: atrioventricular canal defect, CoA: coarctation of aorta, BCPS: bidirectional cavopulmonary shunt, DKS: Damus-Kaye-Stansel Procedure, PA plasty: pulmonary artery plasty, ICR: intra cardiac repair, TCPC: total cavopulmonary connection,

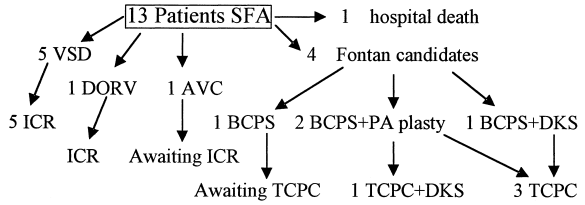

Fig. 3 Flow chart of thirteen study patients on whom subclavian flap aortoplasty (SFA) was performed.

angioplasty. Three patients underwent pulmonary artery plasty in a second operation because of right pulmonary artery stenosis. We performed final operations for five patients who had coarctation with ventricular septal defect and two patients with double outlet right ventricle, and we performed a bidirectional cavopulmonary shunt for four univentricular hearts who are candidates for the Fontan operation. Two patients required Damus-Kaye-Stansel procedure to release restrictive bulboventricular foramen. Three patients underwent a modified Fontan operation (total cavopulmonary connection) after these palliative procedures (**Fig. 3**).

Results

In our series, the intraoperative mortality rate was 0% and post operative mortality rate was 7.7% (1/13). Restenosis occurred in two patients (16.7%) (pressure gradient < 20 mmHg). Ten patients underwent a final operation successfully. A further two patients are considered good candidates for a final operation. The overall mortality was 7.7% (1/13).

Discussion

Although the use of prostaglandin E_1 resuscitation to maintain ductal patency and improve systemic circulation has dramatically improved the preoperative condition of these infants, operative mortality is still a problem[4]. Late mortality may be associated with the progression of pulmonary vascular disease and the presence of associated severe cardiac anomalies. Fontan candidates need staged operations, but there is a marked difficulty in palliative repair for complicated lesions such as transposition of great arteries and truncus arteriosus[5]. Tom R. Karl and colleagues reported that the primary repair of an interrupted aortic arch and a coarctation pulse hypoplastic arch compares favorably with a staged approach and is recommended even when a complex intracardiac anatomy is present[6].

Left ventricular outflow tract obstruction or restric-

tive bulboventricular foramen was present in three of our patients (cases 4, 5, 10). Left ventricular outflow tract obstruction was usually treated by resection through the aorta or the right ventricle. Restrictive bulboventricular foramen, which complicate the clinical course of patients with double-inlet left ventricle and normal relation of great arteries (case 5) or tricuspid atresia and transposition of great arteries (case 4), was usually treated by Damus-Kaye-Stansel procedure[7]. Although it may be difficult to determine whether the bulboventricular foramen is obstructed in neonates because of the patency of the ductus arteriosus, it is well known that a bulboventricular foramen, which is not restrictive in the neonatal period, may become obstructed with time. An attractive management option for left ventricular outflow tract obstruction in the presence of an associated aortic arch obstruction is a Norwood stage 1 procedure[8]. However this procedure can be difficult if pulmonary vascular resistance is increased. Our study shows one case of single atrium, subaortic stenosis pulse hypoplastic arch and intact ventricular septum (case 10) performed as a subclavian flap aortoplasty without pulmonary artery banding who died 3 months after the operation due to the progression of pulmonary hypertension. In our recent pathologic analysis, medial hypertropy of the small pulmonary arteries for coarctation complex demonstrated a more significant progression compared with those of ventricular septal defect without coarctation. Therefore we suspect that the progression of pulmonary vascular disease will be an important factor in the mortality of coarctation complex. Pulmonary vascular disease occasionally progresses even though there is a reduction in the pulmonary arterial pressure just after pulmonary artery banding[9]. One-stage repair would be a reasonable procedure for the patients in whom biventricular repair is feasible, considering the progression of pulmonary vascular disease.

In conclusion, this series of two-stage repair offers a good prognosis for neonates and infants with coarctation complex. Fontan candidates need a staged operation but if biventricular repair is feasible, one-stage repair would be a reasonable procedure considering the distortion of the pulmonary artery due to pulmonary artery banding, progression of pulmonary vascular disease after pulmonary artery banding, and the difficulty in palliative repair such as transposition of great arteries and truncus arteriosus. If an operation can be performed with reasonable risk, it would appear to improve the quality of life of those children.

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