MODIFIED FONTAN PROCEDURE IN NINETY-NINE CASES OF ATRIOVENTRICULAR VALVE REGURGITATION

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Between January 1985 and August 1995, among 242 patients who underwent a modified Fontan procedure, 99 had atrioventricular valve regurgitation ranging in degree from 1 to 4, for which concomitant repair of the atrioventricular valve regurgitation was done in the majority of cases. In all but 4 cases the atrioventricular valve was repaired mainly by circular annuloplasty and valve replacement was not done in any case. Although the hospital mortality rate was significantly higher in cases with atrioventricular valve regurgitation (12/99, 12%) than in cases without (4/143, 3%; $p < 0.0037$, χ^2 test), actuarial survival in atrioventricular valve regurgitation was 84% for years 5 through 10. The degree of atrioventricular valve regurgitation before operation was 1.6 ± 0.7 on average; in 49 cases with higher than grade 2 regurgitation before operation there was a significant decrease to 0.4 ± 0.49 ($p < 0.0001$) after operation in short-term survivors. Patients with atrioventricular valve regurgitation can be treated with reasonable risk, provided proper repair of the valve is done. Circular annuloplasty is a simple and uniformly effective method to control regurgitation even in cases of common atrioventricular valve. (J Thorac Cardio-vasc Surg 1997;113:262-9)

The Fontan procedure was originally used for tricuspid atresia, but its application has been extended to use in patients with a variety of complex cardiac anomalies with a functionally single ventricular chamber. Concomitantly, indication for this operation has been extended further to patients at poor risk. The modified Fontan procedure generally carries higher risk in cases associated with atrioventricular valve regurgitation (AVVR).¹⁻⁶ However, a detailed study on this specific subset has not been reported. The purpose of this presentation was to evaluate parameters before and after the Fontan

procedure in the subset of patients with AVVR, to describe circular annuloplasty, to analyze factors influencing death, and to compare cases with AVVR with those without AVVR.

Material and methods

Between January 1985 and August 1995, among 242 patients who underwent a modified Fontan procedure, 99 patients (41% of the series) had AVVR ranging in degree from 1 to 4, for which concomitant repair of the AVVR was done in the majority of cases. The degree of AVVR (Sellers classification) was evaluated by retrograde cine-ventriculography at preoperative catheterization in all cases and was reevaluated approximately 2 months (60 ± 95.3 days) after the modified Fontan procedure in 78 of 87 short-term survivors. On average, the degree of AVVR was 1.6 ± 0.7 , and 49 cases had higher than grade 2 regurgitation. Those with trivial regurgitation by preoperative Doppler echocardiography were excluded (Table I). Ages at operation ranged from 1 to 27 years (mean 9.95 ± 5.8 years) (Table II), and the male-to-female ratio was 61 to 38. Anomalies consisted of 31 cases of univentricular heart of the right ventricular type, 14 of univentricular heart of the left ventricular type, 7 of classic tricuspid atresia, and 47 other anomalies with biventricular heart including 18 cases with double-outlet right ventricle, 16 with complex atrioventricular (AV) canal defect, 8 with AV discordant anomalies, 4 with complete transposition associated with ventricular imbalance, and 1 with discordant crisscross heart. There were 24 cases of polysplenia and 25 had asplenia syndrome.

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Table I. AVVR group

Sellers classification	Preoperative (No. of cases)	Hospital death (No. of cases)	Postoperative (No. of cases)
Grade 0			52
Grade I	50	5	25
Grade II	38	5	1
Grade III	9	2	0
Grade IV	2	0	0

Before the modified Fontan procedure, 99 palliative procedures had been done in 69 patients consisting mainly of 69 various systemic-to-pulmonary shunt procedures, 10 pulmonary artery banding procedures, open palliative right ventricular outflow reconstruction in 2, and the Norwood operation in 2. Reparative procedures on the valve included circular annuloplasty in 75 cases, conventional annuloplasty in 11, valvuloplasty in 5, patch closure of the valve in 4, and no procedure in 4. Basic morphologic features of the AV valve were common AV valve in 46, two AV valves in 38, and one-sided AV atresia in 15 cases.

Study methods. Catheterization of the right and left sides of the heart with a conventional fluid-filled system and cineangiography in the anteroposterior and lateral projections at a film speed of 60 frames/sec were done in all cases before operation and approximately 2 months (60 ± 95.3 days) after the modified Fontan procedure in 78 of 87 short-term survivors in the AVVR group and in 123 of 139 patients in the non-AVVR group.

Before operation, the volume of the morphologically left ventricle was calculated with the area-length method and the volume of the morphologically right ventricle with Simpson's rule by the method of Graham, Jarmakani, and Canent.⁷ Adjustment of volume occupied by papillary muscles for the left ventricle was made according to the method of Graham and associates⁸ as follows.

When the measured volume of the left ventricle (V) was less than 15 ml, adjusted left ventricular volume was determined by $(V') = 0.874 V - 3.1$, and for left ventricular volume more than 15 ml, the equation $V' = 0.733 V$ was applied.

Similarly, the rudimentary outlet chamber volume of the morphologically right ventricle calculated by Simpson's method was adjusted by the formula $V' = 0.649V$.

The two adjusted ventricular volumes were added to obtain the total volume, whenever two sizable chambers were found, and the total volume was expressed as a percentage of the expected normal ventricular volume per body surface area (BSA) according to the formulas of Nakazawa and associates⁹ as follows:

$$\text{Normal LVEDV} = 72.5 \times \text{BSA}^{1.43}$$

$$\text{Normal RVEDV} = 75.1 \times \text{BSA}^{1.43}$$

Total ejection fraction (VEF) was calculated according to the formula:

$$\text{VEF} = \text{RVEDV} \times \text{RVEF} + \text{LVEDV} \times \text{LVEF/LVEDV} + \text{RVEDV}$$

Table II. Age at operation

Age	No. of cases	Hospital death	Mortality (%)
<4 yr	14	4	28.6
4-15 yr	71	6	8.5
>16 yr	14	2	14.3

Table III. Preoperative parameters

Parameters	AVVR group (range)	Non-AVVR group (range)	p Value
PA index (mm^2/m^2)	366.5 ± 116.5 (213-903)	363.7 ± 1142 (210-1040)	NS
CTR (%)	56.7 ± 7.5 (40-73)	53.9 ± 6.7 (40-72)	<0.003
Mean RAP (mm Hg)	7.9 ± 3.0 (3-23)	8.1 ± 3.4 (2-28)	NS
Mean PAP (mm Hg)	15.9 ± 5.3 (7.5-36)	15.0 ± 4.5 (5-30)	NS
VEDP (mm Hg)	10.2 ± 3.0 (5-20)	9.6 ± 3.0 (4-20)	NS
VEDV (% of normal)	243.3 ± 73.5 (125-536)	216.0 ± 66.1 (74-440)	<0.0033
Ejection fraction (%)	55 ± 6 (36-70)	57 ± 6 (40-76)	NS
PVR (Wood units)	1.66 ± 0.75 (0.49-4.0)	1.86 ± 0.74 (0.26-4.4)	<0.0442
Pao ₂	40.6 ± 5.4 (27.2-55)	39.9 ± 5.8 (26.5-55)	NS

PA, Pulmonary arterial; NS, not significant; CTR, cardiothoracic ratio; RAP, right atrial pressure; PAP, pulmonary artery pressure; VEDP, ventricular end-diastolic pressure; PVR, pulmonary arteriolar resistance index; Pao₂, partial pressure of arterial oxygen.

In these equations LVEDV is the left ventricular end-diastolic volume, RVEDV is the right ventricular end-diastolic volume, RVEF is the right ventricular ejection fraction, and LVEF is the left ventricular ejection fraction.

Data were expressed as a mean value plus or minus one standard deviation and paired *t* test, unpaired *t* test, or analysis of variance was used for data analysis. For discrete factors, contingency table analysis was used and 5% was regarded as the significance level.

Preoperative parameters in the AVVR and non-AVVR groups were tabulated (Table III). Comparison of preoperative parameters in groups with and without AVVR revealed that cardiothoracic ratio ($56.7\% \pm 7.5\%$ vs $53.9\% \pm 6.7\%$; $p < 0.003$) and ventricular end-diastolic volume (VEDV) ($243.3\% \pm 73.5\%$ of normal vs $216.0\% \pm 66.1\%$ of normal; $p < 0.0033$) were significantly larger in the AVVR group, and the pulmonary arteriolar resistance index was higher in the non-AVVR group (1.66 ± 0.75 vs 1.86 ± 0.74 Wood units, $p < 0.0442$). However, differences in pulmonary arterial index,¹⁰ mean right atrial pressure, mean pulmonary arterial pressure, ventricular end-diastolic pressure, ventricular ejection fraction, and arterial oxygen tension remained insignificant.

Fig. 1. Circular annuloplasty. *Upper left*, Gross appearance of common AV valve before repair in an 11-year-old girl with polysplenia, type A complete AV canal with grade 2 regurgitation, small left ventricle, and common atrium. *Upper right*, Purse-string suture was passed around the annulus and, in the vicinity of the conduction system at the posterior junction of the ventricular septum with the AV groove, the suture was passed through the leaflet itself to avoid injury to the conduction tissue. *LSL*, Left superior leaflet; *RSL*, right superior leaflet; *RIL*, right inferior leaflet. *Lower left*, After circular annuloplasty and additional valvuloplasty to close a gap between the superior leaflets, competence was restored. *Lower right*, Purse-string suture was tied.

Operative procedures

Modified Fontan procedure. With the use of general anesthesia and moderate hypothermic extracorporeal circulation (28° to 25° C), direct anastomosis between the atrial appendage and pulmonary artery was done in 83 cases, total cavopulmonary connection in 13 cases, and anastomosis between the right atrium and right ventricle with insertion of a prosthetic gusset (Björk procedure) in 3 cases in our earlier experience.

In a direct appendage–pulmonary artery anastomosis in normal atrial situs, for example, the right atrial appendage is opened along its summit in a figure-of-seven manner, and the distal pulmonary arterial orifice is enlarged by cutting into the right pulmonary artery for 15 to 20 mm to make the largest possible anastomotic orifice. The main pulmonary artery, if present, is always divided and is incorporated in the anastomosis after the appropriate cut into the right pulmonary artery is made to enlarge the anastomotic orifice. The right-sided atrial cavity is

obliquely partitioned toward the root of the appendage with a composite patch of equine pericardium and thin Dacron velour to divert the right-sided AV valve or common AV valve and ostium of the coronary sinus, if present, into the systemic side of the atrium.

Circular annuloplasty. In circular annuloplasty, 2-0 or 3-0 polytetrafluoroethylene suture (CV-2 or CV-3)* is passed through the annulus of the AV valve circumferentially and through the leaflet itself in the vicinity of the conduction tissue to avoid conduction disturbance, and then the full circular purse-string suture is tied over a Hegar's dilator with a diameter from 22 to 24 mm (Fig. 1). In a biventricular heart with concordant AV relationship, the dangerous area of AV conduction can be easily recognized as the Koch's triangle, and a bite is taken in the

*Gore-Tex suture, registered trade name of W. L. Gore & Associates, Inc.

Table IV. Types of valve repair and result

Type of repair	No. of cases	Hospital death	Late death
Circular annuloplasty	75	10	4
Conventional annuloplasty	11	2	0
Valvuloplasty	5	0	0
Valve closure	4	0	0
None	4	0	0

medial leaflet of the tricuspid valve. In complex heart anomalies without formation of the coronary sinus, the suture is applied to the leaflet itself to avoid injury in the vicinity of the point of the junction of the ventricular septum with the posterior AV groove. In cases with discordant AV relationship including univentricular heart of the left ventricular type, the AV ring at the insertion of the interventricular septum can be recognized as the site of anterior conduction. In univentricular hearts of the right ventricular type with recognizable rudimentary left ventricle, both the anterior and posterior extremities of the ventricular septum that connect with the AV groove are considered as dangerous areas, and in those hearts without an identifiable rudimentary chamber, the posterior AV valve ring should be avoided to prevent injury. This method was most frequently applied in this series. Unlike conventional annuloplasty uniform constriction of the AV valve after full-circular annuloplasty can provide maintenance of the original composition of each leaflet and effectively reduce the orifice area. In the modified Fontan procedure drastic reduction in total cardiac output after the operation allows for drastic reduction of the AV valve orifice area by more than one half of the original area.

Other reparative procedures. In the presence of a cleft, simple suture closure of the cleft was done. Conventional annuloplasty of the Kay or Reed type was applied for regurgitation limited to one commissure. In cases with two AV valves, of which the diminutive one was regurgitant, patch closure of the valve was done.

Results

The hospital mortality rate was significantly higher in cases with AVVR (12/99, 12%) than in cases without AVVR (4/143, 3%; $p < 0.0037$, χ^2 test). A summary of results by type of valve repair is given in Table IV. Four patients died late during a mean follow-up period of 4.3 ± 2.9 years and ranging from 2 to 124 months. Actuarial survival by the Kaplan-Meier method revealed 84% survival from 1 through 10 years for the AVVR group and 95% at 5 years and 93% at 10 years for the non-AVVR group (Fig. 2). Late deaths occurred in 4 cases at 2, 7, 7, and 11 months, respectively. Causes of late death were systemic ventricular failure in 3 and massive melena caused by midgut volvulus in 1 patient with asplenia syndrome. Reop-

Fig. 2. Actuarial survival by the Kaplan-Meier method.

eration during the same hospitalization was required in 11 patients after the modified Fontan procedure. Takedown of the Fontan repair was done in 5 cases because of severe low cardiac output state and all of these patients died. The other 7 hospital deaths occurred 22.7 ± 17.3 days after operation with a range from 8 to 49 days. Causes of these 7 deaths were renal failure in 3, respiratory failure in 1, pneumonia in 1, hypoxia because of bronchial bleeding in 1, and massive gastrointestinal tract bleeding in 1. Repeat valvuloplasty was required in 3 cases. In 2 patients circumferential annuloplasty resulted in laceration of the leaflet in the vicinity of the conduction system and repair of the leaflet was done. In 1 patient persistent hemolysis necessitated reintervention and patch closure of a leaking commissure was done. All three of these patients survived. Repair of residual interatrial shunt was done in 3 patients and 1 died. Regarding conduction disturbance, new complete AV block occurred in 2 cases after operation. Sixty-nine (75%) of 99 patients have regular sinus rhythm.

The degree of AVVR decreased significantly from 1.64 ± 0.74 to 0.4 ± 0.49 ($p < 0.0001$) after operation in 78 of 87 short-term survivors (Table I). In the remaining 9 cases in which AVVR was evaluated only by Doppler echocardiography, 5 showed no or trivial regurgitation, 2 mild regurgitation, 1 mild to moderate regurgitation, and 1 massive regurgitation. The latter patient, a 25-year-old

Table V. Postoperative parameters

Parameters	AVVR group (range)	Non-AVVR group (range)	p Value
PA index (mm ² /m ²)	266.5 ± 99.9 (125-591)	264.5 ± 93.7 (111-590)	NS
CTR (%)	56.5 ± 6.9 (37-74)	55.9 ± 6.5 (39-71)	NS
Mean RAP (mm Hg)	14.3 ± 3.2 (7-24)	13.4 ± 3.0 (5-20)	<0.0404
Mean PAP (mm Hg)	14.0 ± 3.3 (6-23)	12.7 ± 3.0 (4-20)	<0.0059
VEDP (mm Hg)	6.9 ± 2.8 (2-16)	6.7 ± 3.0 (1-17)	NS
VEDV (% of normal)	152.4 ± 49.7 (48-327)	136.6 ± 42.2 (40-301)	<0.0205
Ejection fraction (%)	44 ± 11 (17-66)	48 ± 9 (29-71)	<0.017
PVR (Wood units)	2.5 ± 0.9 (0.9-5.5)	2.2 ± 1.1 (0.3-5.7)	NS
PaO ₂ (mm Hg)	57.6 ± 9.6 (42-93)	61.3 ± 10.4 (38-98)	<0.0133

PA, Pulmonary arterial; NS, not significant; CTR, cardiothoracic ratio; RAP, right atrial pressure; PAP, pulmonary artery pressure; VEDP, ventricular end-diastolic pressure; PVR, pulmonary arteriolar resistance index; PaO₂, partial pressure of arterial oxygen.

Table VI. Diagnoses

Diagnosis	No. of cases	Hospital death		Late death
		No.	%	
TA	7	3	42.8	0
UVH-RV	31	2	6.5	2
UVH-LV	14	0	0	1
BV	47	7	14.9	1

TA, Classic tricuspid atresia; UVH-RV, univentricular heart of right ventricular type; UVH-LV, univentricular heart of left ventricular type; BV, other complex biventricular heart anomalies.

man with corrected transposition, mitral atresia, and massive tricuspid regurgitation, underwent reoperation on the first postoperative day because of failed circular annuloplasty as a result of laceration of a leaflet and is alive with residual regurgitation 54 months after operation. No valve repair was done in 4 patients despite preoperative grade 1 AVVR, because there was no apparent jet lesion on or around the valve on inspection. In these 4 cases the degree of AVVR remained the same after operation, and all patients survived. Postoperative pulmonary wedge pressures were the same in the groups with and without preoperative regurgitation: 7.9 ± 2.9 mm Hg in the AVVR group and 7.3 ± 2.4 mm Hg in the non-AVVR group ($p = 0.3646$). Rates of reduction in VEDV after operation were also the same in both groups: 66.2% ± 19.9% of the preop-

Table VII. Basic morphologic types of AV valve

AV valve morphologic type	No. of cases	Hospital death		Late death
		No.	%	
Common	46	6	13.0	3
Two	38	3	7.9	1
Atresia	15	3	20	0

Common, Common AV valve; two, two AV valves; atresia, one-sided valvular atresia.

erative volume in the AVVR group and 66.3% ± 20.1% in the non-AVVR group ($p = 0.9681$). Rates of reduction in ejection fraction after the modified Fontan procedure were, however, significantly larger in the AVVR group: 79% ± 19% of the preoperative value with AVVR versus 85% ± 15% without AVVR ($p < 0.0185$) (Tables III and V).

Preoperative factors that influenced hospital death and survival were the number of parameters that exceeded the limits of Choussat's 10 commandments excluding right atrial size 4.92 versus 3.83 ($p < 0.0077$), cardiothoracic ratio 62% versus 56% ($p < 0.0046$), VEDV 283% versus 236% of normal ($p < 0.0385$), pulmonary blood flow 3.5 versus 5.0 L/min per square meter ($p < 0.0202$), and Mayo's criterion (pulmonary vascular resistance + ventricular end-diastolic pressure/pulmonary flow + systemic flow) 3.74 versus 2.95 ($p < 0.0248$). However, ventricular ejection fraction, pulmonary vascular resistance, degree of AVVR 1.75 versus 1.62, morphologic type of systemic ventricle, and ventricular end-diastolic pressure were not related to death.

The basic diagnoses including tricuspid atresia, univentricular heart of the right ventricular type, univentricular heart of the left ventricular type, and biventricular heart had a statistically significant effect on hospital death in the AVVR group ($p < 0.0318$) (Table VI), but this was not significant in the entire series ($p = 0.5277$, χ^2 test). The morphologic type of AV valve; whether there was common, double, or one-sided atresia ($p = 0.4742$); and the presence or absence of heterotaxia ($p = 0.685$) were not related to hospital death (Tables VII and VIII). The factor that influenced late death in the AVVR group was postoperative ejection fraction: 45% ± 10% in 70 survivors and 22% ± 5% in 3 late deaths studied ($p < 0.0001$).

Discussion

In the modified Fontan procedure, the presence of AVVR is regarded as one of the risk factors²; however, only a few reports dealing specifically with this problem in large series have been published,³⁻⁶

Table VIII. Heterotaxia

Heterotaxia	No. of cases	Hospital death		Late death
		No.	%	
Polysplenia	24	4	16.7	0
Normal	50	6	12.0	3
Asplenia	25	2	8.0	1

and in these the mortality rate ranged from 15% to 60%.³⁻⁶ AVVR in complex heart anomalies is quite common especially in those associated with heterotaxia. The incidence of AVVR in heterotaxic hearts was more than twice that in nonheterotaxic hearts in this series: 69% (49/71 cases) versus 29.2% (50/171 cases) ($p < 0.0001$). The incidence of AVVR is influenced by the morphologic type of the AV valves. The highest incidence is observed in cases of common AV valve, the second highest in cases with two AV valves, and the lowest in cases with one-sided AV valve atresia (73%, 39%, and 19%, respectively). Thus cases with heterotaxia or with common AV valve, or both, tend to have the highest incidence of regurgitation. The natural course of AVVR is poor¹¹ and in our series patients with these complex anomalies had marked dilation and thinning of the ventricular wall with resultant depressed ejection fraction in a matter of a few to several years. The incidence of AVVR was relatively high in our series, because our policy was to achieve the best possible pulmonary vascular bed before the modified Fontan procedure by increasing pulmonary blood flow by application of systemic-to-pulmonary shunts. The preoperative pulmonary blood flow index (Qp) inversely affected surgical outcome in our entire series: 3.2 ± 1.2 L/m² in patients with hospital death and 4.5 ± 2.2 L/m² in survivors ($p < 0.0334$). Although the difference was statistically insignificant ($p = 0.1481$), the hospital mortality rate was lower in the group with higher Qp (>4 L/m²) than in the group with lower Qp (<4 L/m²): 3.6% (4/107) versus 8.2% (9/101).

One of the characteristic features of Fontan circulation is a drastic reduction in total cardiac output after operation. The preoperative total cardiac index including both systemic (Qs) and pulmonary blood flows (Qp + Qs) averaged 8.51 ± 2.53 L/min per square meter, whereas it became one third of the preoperative value, 2.75 ± 0.68 L/min per square meter, after operation ($p < 0.0001$). Therefore the orifice area of the AV valve can be drastically reduced to 32% of the preoperative value. Thus the postoperative diameter of the valve can be

Fig. 3. Ventricular ejection fraction. VEF, Preoperative ventricular ejection fraction; VEF-PI, postoperative ventricular ejection fraction.

as small as 58% of the preoperative value even disregarding regurgitant fraction, and this accounts for the effectiveness of circular annuloplasty in this series. This drastic reduction in flow through the AV valve after operation should reduce VEDV and this also favors reduction of AVVR after a modified Fontan repair even without repair of the AV valve. Theoretically, postoperative VEDV should be 32% of the preoperative volume, if the ejection fraction remains unchanged. However, postoperative VEDV was only 66% of the preoperative value 60 days after operation and the ejection fraction also decreased to $79\% \pm 18\%$ of the preoperative value in this series. Therefore appropriate surgical measures to reduce annular size seemed mandatory.

Meticulous and time-consuming valvuloplasty with appropriate annuloplasty can be a better measure to control valvular regurgitation in normal circumstances. However, in the modified Fontan procedure for complex cardiac anomalies, complexity and diversity in the anatomy of the AV valve make standardized repair infeasible; moreover, shortening of cardiopulmonary bypass time is important to maintain compliance in the lung for a better hemodynamic state in the immediate postoperative period. Also, the cause of AVVR is mainly annular enlargement or prolapse of leaflets. Even a grossly malformed valve has restored competence if the annular diameter is reduced to nearly 60% of the original diameter. The circular annuloplasty is also effective to control regurgitation in cases of common AV valve. Laceration of a leaflet in the vicinity of the conduction system is a disadvantage of this circular annuloplasty technique and was seen in two of our patients. Reinforcement of leaflet tissue

with autologous pericardial patch may reduce the incidence of disruption of the leaflet. Another possible demerit is conduction disturbance. However, this can be avoided by proper identification of the conduction system in normal circumstances, and new complete AV block was seen in only 2 of our 99 cases.

In rare instances, a small anomalous leaflet is completely prolapsed into the atrial cavity as a result of marked elongation or rupture of chordae. The prolapsed leaflet can be approximated with adjoining leaflets with a running over-and-over suture over an autologous pericardial tape before circular annuloplasty.

That reduction in ejection fraction was larger in the AVVR group than that in the non-AVVR group may indicate reduced ventricular function in this group. AVVR tends to induce progressive passive dilation of ventricle unaccompanied by appropriate hypertrophy because of the absence of isometric contraction. This resultant dilated ventricle with a relatively thin wall accounts for the depressed ejection fraction after repair of the regurgitation by a sudden increase in afterload. VEDV values before and after operation are significantly correlated in both groups ($Y = 0.38X \pm 58.7$, $r = 0.59$, $p < 0.0001$, where Y is postoperative VEDV as a percent of normal and X is preoperative VEDV). Thus a larger ventricle undergoes a larger reduction rate in volume after operation, and theoretically the ejection fraction is reduced to a greater extent in larger ventricle. However, the ratio of ejection fraction after and before a modified Fontan procedure in the larger ventricle group (VEDV $>250\%$ of normal) was significantly smaller in the AVVR group than that in the non-AVVR group: $74\% \pm 19\%$ of preoperative value versus $84\% \pm 17\%$ ($p = 0.0412$). Therefore it is predicted that the postoperative ejection fraction will be reduced by 26% in the AVVR group, whereas in the non-AVVR group its reduction will be only 16%.

Changes in ejection fraction after operation are, however, quite unpredictable (Fig. 3). A drastic reduction in ejection fraction was only observed in some cases in the AVVR group. A low ejection fraction less than 40% after operation was observed in 24 cases in the AVVR group; however, the ejection fractions in these patients before operation averaged $54\% \pm 6\%$ and ranged from 37% to 70%. Low ejection fraction before operation usually remains similar or even shows slight improvement after operation. Therefore low ejection fraction itself does not preclude performance of a modified

Fontan procedure. However, postoperative low ejection fraction constitutes a definite risk factor of late death in the AVVR group.

The mortality rate in the AVVR group was almost three times higher than that in the non-AVVR group in this series; however, a favorable hemodynamic state can be achieved after operation in the majority of survivors provided reasonable repair of the regurgitation is done. Actuarial survival in the AVVR group was lower than that in the non-AVVR group; however, 84% survival in years 5 through 10 in the former group is quite acceptable. Circular annuloplasty is a simple and uniformly effective method to control regurgitation even for cases with common AV valve.

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Discussion

Dr. Hillel Laks (*Los Angeles, Calif.*). Dr. Imai and his colleagues have described a group of patients with challenging cardiac disease who underwent the Fontan procedure, and they have shown that the group of patients with AVVR had an increased early mortality rate even though the majority of these patients had grade 1 to 2 regurgitation. This excellent series with a mortality rate of only 3% in the nonregurgitant group raises several important questions.

The first is the question of staged treatment in patients with moderate and particularly severe degrees of regurgitation. It has been our practice to stage treatment in such patients with severe regurgitation, particularly if they have ventricular dilation or dysfunction, with an initial bidirectional Glenn shunt combined with a valve repair. Only 11 of the patients in this series, I think, had grade 3 or 4 regurgitation and none had preoperative Glenn shunts. Do the authors select patients for a staged approach, and what are their criteria with regard to the degree of regurgitation, the hemodynamics, and the ventricular function?

I noticed that age was a factor in mortality and that patients younger than 4 years old had a 28% mortality rate. What accounts for this increased mortality, and does it influence the authors' attitude or position with regard to staging in the younger patients?

The technique that has been described is that of a circumferential annuloplasty with use of a polypropylene suture and reduction of the circumference quite drastically to 60% of the previous diameter. The conduction tissue is avoided by placement of sutures at the base of the valve leaflets. I noticed in the manuscript that there was, I believe, one patient in whom the valve leaflet tissue tore. We have found that the polypropylene suture annuloplasty used in other circumstances and in AV canals has a tendency to unravel with time; in some patients, dependent on the annuloplasty, late regurgitation has recurred. We have therefore preferred the use of a pericardial strip annuloplasty done in several segments, avoiding the area of the conduction tissue where the annuloplasty is interrupted, and we have begun to use polytetrafluoroethylene sutures as a suture in which tissue ingrowth occurs, which will possibly prevent the unraveling. Have the authors seen late failure with their annuloplasties, particularly in the group with grade 3 or 4 regurgitation? Have they documented growth of the annuloplasty, particularly in the smaller children, or are they concerned that this relatively tight annuloplasty could result in a lack of growth in the future?

The mortality rate in the common AV valve group was higher in this series. Was this related to residual AVVR? In the AV canal type of common AV valve with moderate to severe regurgitation, particularly of long standing, in which one may find elongation of chordae and distortion of the valve leaflets, we have found that an annuloplasty alone in the severely regurgitant group is frequently not adequate. We have found that we have had to add a central support to the valve by the suturing of clefts in the neomitral part of the valve and by suturing the base of the tricuspid component of the valve, and with this central support and the annuloplasty we have achieved competence. Do the authors have any comments on this particular subgroup?

The early mortality rate was elevated in the regurgitant

group. Do the authors have any experience with the adjustable atrial septal defect or fenestration? Although this fenestration or adjustable atrial septal defect technique was initially described in particular for high transpulmonary gradients, we have also found it useful in patients with elevated left atrial pressure. Do the authors have any experience in this particular interesting group of patients with regurgitation?

It is promising that so far the long-term results of the combined procedure are similar to those of the nonregurgitant group, at least to the 5 or 10 years of follow-up that the authors so far have. However, that the ejection fraction, the end-diastolic volume, and the right atrial pressure are statistically higher in this group than in the nonregurgitant group gives us concern about the 10-, 15-, and 20-year follow-up for these patients. It suggests that we might well avoid volume-loading shunts in these patients, and it suggests that we should move to a non-volume-loading bidirectional Glenn shunt very early, in the first 3 to 6 months of life, particularly in the single ventricle group. I would like to know what the authors' opinion is about early performance of the Glenn procedure for these patients.

In conclusion, Dr. Imai and his colleagues are to be congratulated for superb results in their Fontan operation series and for the technical innovations that they have proposed for the regurgitant valves, as for the innovations they have made in the treatment of many other types of complex congenital lesions.

Dr. Imai. Thank you very much, Dr. Laks. Regarding the staging procedure, this series dates back almost 10 years. In our initial experience, we did the Kawashima operation in the presence of AVVR, but the rate of reduction in preload in this type of palliation was not as much as that seen in the modified Fontan repair. Therefore in repairing the valve, even if circular annuloplasty or any kind of annuloplasty is used, one cannot reduce the size of the valve efficiently enough in the presence of a bidirectional Glenn or Kawashima-type procedure. That is the reason we like to repair the valve concomitantly with the Fontan-type repair.

As to the question regarding circular annuloplasty, we usually use polytetrafluoroethylene suture to restrict the annulus evenly. There were three cases in which we had to reoperate because of residual regurgitation. In two, there was a disruption of the leaflet. In these cases, we reinforced the lacerated portion of the leaflet with autologous pericardium and were able to successfully control the residual regurgitation. Especially when the leaflet is thin and tissue integrity is poor, such a reinforcing measure has to be taken before a bite can be taken in the leaflet itself.

Regarding early intervention with performance of a bidirectional Glenn procedure at an early age, the majority of the cases in our series did have pulmonic stenosis or atresia in low-flow situations with a small pulmonary artery to begin with. That was the reason we needed to develop a sizable pulmonary artery by putting in a shunt. If there was a high-flow situation, as Dr. Laks mentioned, probably staging in early infancy would reduce the mortality rate. However, in our recent series of the past 5 years, there was no difference in the mortality rate whether the patient was younger than 4 years old or between 5 and 15 years.