Impact of Fontan Operation on Left Ventricular Size and Contractility in Tricuspid Atresia

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Left ventricular dimensions and contractility were determined by echocardiography in 33 patients with tricuspid atresia in 1985 and again in 1988. Eight patients remained palliated throughout the 3-year period; neither the left ventricular end-diastolic diameter (153±15% of normal vs. 157±19%, p=NS) nor a load-independent index of contractility (rate-corrected velocity of shortening [VCFc]/end-systolic meridional stress [ESSM]) changed. Eleven patients underwent a Fontan operation during the study and were reevaluated at least 6 months after surgery; left ventricular dimension decreased (130±15% vs. 114±19%, $p<0.001$), and the contractility index VCFc/ESSM improved ($p<0.05$). Fourteen patients had undergone a Fontan operation 0.9–9.5 years (mean, 4.2 years) before initial examination in 1985. Over the 3-year period, left ventricular dimensions did not change (121±17% vs. 118±11%, p=NS), but the contractility index showed significant improvement ($p<0.01$). Eight additional patients were studied just before and after a Fontan operation to examine the early effects of surgery. Left ventricular dimensions decreased from 130±14% to 100±13% by 10 days $p<0.001$) with no further change at 2 months. An inappropriate degree of ventricular hypertrophy was observed in only the early postoperative period. Successful Fontan repair results in rapid reduction of left ventricular size, followed by regression of hypertrophy to a normal mass-to-volume ratio. Operating at more favorable dimensions and loading conditions results in an early increase in left ventricular contractility, which further improves in the medium term follow-up. (*Circulation* 1990;81:118–127)

There have been numerous reports of abnormal left ventricular dimensions and function in patients with tricuspid atresia, both before and after a Fontan operation.[1-12] Various factors might contribute to these morphologic and functional abnormalities, including preoperative chronic volume overload and cyanosis, perioperative myocardial damage, and adverse postoperative hemodynamics. All previous studies, however, have measured left ventricular pump function in terms of variables that are affected not only by abnormal contractility but also by afterload, preload, and myocardial hypertrophy.

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Colan et al[13] reported an echocardiographically derived index of left ventricular function that is a sensitive measure of contractile state and is relatively independent of abnormal loading conditions. In 1985, we reported a cross-sectional study using this index to compare left ventricular function in patients palliated for tricuspid atresia with those who had undergone a Fontan operation.[14] Reexamining these patients after a 3-year period, we now report a prospective longitudinal study that reports the effect of the altered loading conditions on left ventricular size and function after a Fontan operation. Our aim was to understand whether the Fontan operation represents a beneficial long-term investment for ventricular function in patients with tricuspid atresia.

Methods

In 1985, we studied all patients with tricuspid atresia who underwent surgery (palliation or Fontan procedure) at The Hospital for Sick Children, London, from January 1970 to January 1985 and were

living in the UK. Patients were excluded if they were less than 18 months old or had transposed great arteries with subvalvar aortic stenosis. Four additional patients with pulmonary atresia and intact ventricular septum, severe tricuspid stenosis, and diminutive right ventricles were also included as their hemodynamics were believed to be functionally similar to tricuspid atresia. Of the total of 42 patients studied in 1985, 33 were reevaluated in 1988 (study interval, 2.7–3.1 years; mean, 2.8 years). Four patients died during the study period (two early and two late after a Fontan procedure). The remaining five patients declined reevaluation for nonmedical reasons, but all are known to be alive and well.

All patients were studied electively and were considered to be in stable clinical condition. No patient had edema, effusions, or protein losing enteropathy; all had a mild degree of hepatomegaly. The 33 patients were divided into three groups. Group 1 consisted of eight patients who remained palliated throughout the study period. Since the initial evaluation, four shunt procedures were performed in three patients. Five patients are considered potential candidates for a Fontan repair; the other three have pulmonary vascular problems (pulmonary artery hypoplasia in one and severe distortion in two) (Table 1).

Group 2 consisted of 11 patients who had a Fontan operation by direct right atrial-pulmonary artery connection between 1985 and 1988. During this period, only patients who were increasingly symptomatic, typically from cyanosis, were selected for a Fontan operation. All patients were studied before and at least 6 months after Fontan repair (Table 2).

The 14 patients in group 3 had undergone a Fontan operation 0.9–9.5 years (mean, 4.2 years) before initial examination in 1985. Longitudinal comparison was, therefore, available between two postoperative studies (Table 3).

To examine the early effects of the Fontan operation on left ventricular function and size, a fourth group (group 4) of eight consecutive patients with tricuspid atresia undergoing the Fontan operation between September 1987 and April 1988 was investigated. Echocardiographic examination was performed just before surgery and 10 days and 2 months after the Fontan procedure. All patients in this group were taking digitalis, diuretics, and vasodilators at the first postoperative evaluation; at 2 months, medication in all patients had significantly been reduced (Table 4).

Echocardiographic Studies

All patients underwent echocardiographic examination with an Advanced Technology Laboratory Mark 600 Ultrasound System in 1985 and with an Ultramark 8 in 1988. Studies were performed with patients supine or in a slight left lateral decubitus position in a darkened, quiet room, with sedation required in a few young patients in 1985. Mitral regurgitation, residual ventricular septal defect, subaortic stenosis, and aortic regurgitation were excluded in all patients by Doppler echocardiographic analysis.

Precordial long- and short-axis views of the left ventricle were used to detect regional wall motion abnormalities; two patients in 1985 and one patient in 1988 were excluded because of marked septal dyskinesia. Simultaneous recordings of left ventricular echocardiogram, phonocardiogram, indirect carotid pulse tracing, and electrocardiogram were obtained. Systolic and diastolic blood pressures were recorded with a cuff sphygmomanometer.

Measurements and Calculations

Left ventricular internal dimensions (D) and posterior wall thickness (h) were measured at end diastole (ed) and end systole (es) for five beats, and mean values were computed. Ed was defined as the onset of the QRS, and es was estimated by the first high-frequency component of the second heart sound. Left ventricular ejection time (ET) was measured from the carotid pulse tracing and rate-corrected to a heart rate of 60 beats/min by dividing by the square root of the R-R interval (ETc). Es pressure (P_{es}) was estimated by the method of Borow et al[15] by assigning maximum systolic blood pressure to the peak and diastolic pressure to the nadir of the carotid pulse, with subsequent linear interpolation to the level of the incisura. Fractional shortening (FS) was calculated as (Ded−Des)/Ded, and endocardial velocity of circumferential fiber shortening corrected for heart rate (VCFc) was determined as FS/ETc. Endocardial es meridional stress (ESSM) was determined by the method of Grossman et al[16] in which ESSM (g/cm^2) = $Pes \times Des \times 1.35/4 \times h_{es}(1+h_{ed}/D_{es})$ where D_{es} and h_{es} are in centimeters, P_{es} in millimeters of mercury, 1.35 is a conversion factor (from mm Hg to g/cm^2), and 4 is a geometric factor that results from conversion of radius to internal diameter.

Normal values for D_{ed} were estimated as 37.75+12.88 log (body surface area [BSA]) from the study of Gutgesell et al.[17] Dimensions and contractility indexes were compared with literature data and our own results from 42 normal controls studied in our laboratory.[18]

Statistical Analysis

Data were stored in a SIR (version 2) database on an Amdahl mainframe computer and analyzed using SAS statistical packages (version 10, Northwestern University). A Shapiro-Wilk normality test was performed on the differences of paired data; the significance level was set at 0.10. Depending on the normality of the distribution, the *t* test for paired data or the Wilcoxon ranked sum test was used; *p* values obtained by the latter test are identified in the tables. Associations between variables were analyzed with the Pearson product-moment correlation coefficient. A *p* value of less than 0.05 was considered statistically significant. All data are expressed as mean±SD.

TABLE 1. Patients With Palliation Only (Group 1)

Patient	Age in 1988 (yr)	Previous surgery age (yr)	Surgery after 1985	BSA 1985 (m^2)	BSA 1988 (m^2)	Ded 1985		Ded 1988		hed 1985		
						mm	%BSA	mm	%BSA	mm	%BSA	%Ded
1	8.1	BT 0.1, BT 0.1, CS 2.4		0.687	0.893	67.2	188	59.2	161	6.4	119	56
2	13.3	WAT 0.2, BT 8.0		0.842	0.931	59.4	161	65.1	174	8.1	138	81
3	5.3	Band 0.4		0.511	0.775	48.5	142	47.6	131	6.2	130	76
4	6.4	BT 0.01, BT 1.7	BT 4.5	0.661	0.837	48.2	136	54.2	147	6.5	123	80
5	6.8	BT 0.02		0.528	0.676	61.5	180	64.5	181	9.2	191	89
6	17.9	WAT 0.03, BT 8.3, BT 10.3	CS 16.0	1.34	1.46	64.8	164	73.6	185	7.4	103	67
7	7.4	CS 0.01, BT 1.7	CS 6.0, CS 6.7	0.627	0.817	42.8	122	50.9	139	6.3	121	87
8	9.0	BT 0.3, BT 3.0		0.645	0.798	47.2	134	52.3	143	5.7	108	71
Mean	9.3			0.730	0.898	54.9	153	58.4	157	6.9	129	76
±SD	4.2			0.267	0.240	9.3	15	8.8	19	1.2	27	11
p					<0.001			NS	NS			

Band, pulmonary artery band; BSA, body surface area; %BSA, % of normal for BSA; BT, Blalock-Taussig shunt (classic or modified); CS, central shunt; Ded, ventricular end-diastolic dimension; %Ded, % of normal for Ded; G, glycosides; hed, ventricular end-diastolic posterior wall thickness; NYHA, New York Heart Association; WAT, Waterston shunt.
All *p* values refer to a paired *t* test unless stated otherwise.

Interobserver and intraobserver variability were assessed for all variables for 30 cardiac cycles in five patients. Values for the coefficients of variation ranged between 0.2% and 4.9% for interobserver variability and between 0.3% and 4.2% for intraobserver variability.

Results

Dimensions

Although left ventricular end-diastolic dimensions increased in six of the eight patients who remained palliated throughout the study period (group 1), the overall change for the group did not reach statistical significance (153±15% of normal vs. 157±19% of normal, p=NS) (Figure 1). In contrast, the end-diastolic dimension of patients having received a Fontan repair during the study period (group 2) decreased significantly both in absolute values (−9±8%, $p<0.02$) and when expressed as percentage of normal (130±15% vs. 114±19%, $p<0.001$). Only two patients in group 2 showed no decrease in ventricular size. One of these patients had developed

TABLE 2. Patients With Fontan Repair During Study Period 1985–1988 (Group 2)

Patient	Age in 1988 (yr)	Previous surgery age (yr)	Age at Fontan (yr)	BSA 1985 (m^2)	BSA 1988 (m^2)	Ded 1985		Ded 1988		hed 1985			hed 1988
						mm	%BSA	mm	%BSA	mm	%BSA	%Ded	mm
1	4.6	BT 0.3, BT 0.3	3.7	0.495	0.713	34.3	188	33.3	161	7.5	160	129	5.2
2	5.0	BT 0.2, WAT 1.8	2.6	0.491	0.690	40.3	119	37.8	106	6.6	141	97	5.5
3	5.6	BT 0.9, BT 2.0	3.8	0.519	0.728	46.7	137	41.6	116	6.3	132	80	5.9
4	5.7	BT 1.5	4.8	0.507	0.713	43.2	127	35.8	100	6.5	137	89	6.5
5	6.9	BT 0.1, BT 4.6	6.3	0.661	0.906	48.2	136	42.4	114	6.5	122	80	8.0
6	7.1	BT 3.5	4.9	0.594	0.846	41.3	119	38.4	104	6.5	128	93	6.4
7	9.4	BT 0.01, BT 3.6	8.0	0.841	1.140	57.1	155	52.0	135	6.4	109	66	7.0
8	9.5	BT 0.02, BT 3.6	8.3	0.784	1.010	47.5	131	39.8	105	8.1	142	101	6.8
9	9.8	BT 0.1, BT 1.0	8.0	0.759	0.900	46.1	127	38.5	103	5.8	103	74	6.4
10	11.7	BT 0.01, BT 6.2	11.1	0.809	1.050	47.5	130	48.0	126	6.7	116	83	8.3
11	13.6	BT 2.2, CS 7.6	11.3	0.905	1.180	57.5	154	62.3	161	8.2	135	84	6.7
Mean	8.1		6.6	0.670	0.898	46.3	130	42.7	114	6.8	130	89	6.6
±SD	3.0		3.0	0.156	0.165	6.8	15	8.4	19	0.7	16	17	0.9
p					<0.001			<0.05	<0.001				NS

BSA, body surface area; %BSA, % of normal for BSA; BT, Blalock-Taussig shunt (classic or modified); CS, central shunt; Ded, ventricular end-diastolic dimension; %Ded, % of normal for Ded; G, glycosides; hed, ventricular end-diastolic posterior wall thickness; NYHA, New York Heart Association; WAT, Waterston shunt; D, diuretics; V, vasodilator (angiotensin converting enzyme inhibitor).
All *p* values refer to a paired *t* test unless stated otherwise.

TABLE 1. *(Continued)*

hed 1988			VCFc (cycles/sec)		ESSM (g/cm²)		Medi- cation	NYHA class
mm	%BSA	%Ded	1985	1988	1985	1988	1988	1988
6.1	101	61	0.656	0.760	115	103	G	III
8.0	130	73	0.881	0.910	79.6	50.5	G	II
6.8	120	85	1.117	1.020	51.9	38.9	G	II
7.0	119	76	1.030	1.030	52.4	52.0	–	II
8.9	166	82	0.942	1.000	63.1	46.6	–	II
8.2	110	66	0.790	0.833	81.2	68.9	–	II
7.5	129	87	1.040	1.110	49.2	44.8	–	III
7.5	130	85	0.849	0.863	47.7	56.9	–	II
7.5	126	77	0.916	0.928	67.5	57.7		
0.9	19	8	0.153	0.104	23.3	20.4		
NS	NS	NS		NS		<0.05		

asymptomatic atrial flutter between 14 and 22 months after the Fontan operation with a ventricular rate of 110–140 beats/min on a 24-hour Holter monitor. A postoperative echocardiogram in atrial flutter 22 months after the Fontan operation showed a substantial increase in end-diastolic dimension. With diuretics and control of the ventricular rate with digitalis, this improved but still remained above preoperative values. In group 2, there was a highly significant correlation between end-systolic and end-diastolic left ventricular absolute dimensions before the Fontan procedure and the postoperative end-diastolic measurements (r=0.92, p<0.001; r=0.88, p<0.001, respectively).

There was no significant change in end-diastolic dimension in group 3 patients in whom both measurements were made during follow-up after the Fontan procedure (121±17% vs. 118±11%, p=NS).

In the eight group 4 patients whose dimensions were measured immediately before and early after the Fontan procedure, there was a significant decrease in end-diastolic dimension (130±14% vs. 100±13%, p<0.001). There was no further change at the 2-month evaluation (101±12%, p=NS). The variable pattern of end-diastolic dimension during both evaluations may reflect differences in medical therapy. As in group 2, the postoperative end-diastolic dimension at 10 days correlated best with preoperative end-systolic dimensions (r=0.91, p<0.002) but also with end-diastolic dimensions (r=0.89, p<0.003).

TABLE 2. *(Continued)*

hed 1988		VCFc (cycles/sec)		ESSM (g/cm²)		Medi- cation	NYHA class
%BSA	%Ded	1985	1988	1985	1988	1988	1988
95	92	1.030	1.050	37.5	37.1	–	I
102	86	1.071	1.090	37.2	54.5	–	I
106	84	0.701	0.560	53.0	68.3	–	I
118	108	1.030	1.162	44.0	52.7	–	I
132	112	1.048	1.291	52.4	28.7	–	I
108	99	1.020	1.047	47.9	51.9	–	I
104	80	0.710	0.963	76.2	73.3	–	I
107	101	0.966	0.997	31.6	44.6	–	I
105	98	1.061	1.496	52.9	30.0	–	I
128	102	0.770	1.036	63.0	43.0	GDV	II
98	64	0.811	0.899	57.5	62.3	GD	II
110	94	0.929	1.054	50.3	49.2		
12	14	0.149	0.233	12.8	15.4		
<0.02	NS		<0.05		NS		

Wall Thickness

Left ventricular wall thickness did not change significantly in patients who remained palliated (group 1) (Figure 2). It was increased when compared with normal values for BSA but was decreased when related to end-diastolic dimension (D_{ed}), suggesting ventricular dilatation (BSA: 129±27% vs. 126±19%, p=NS; D_{ed}: 76±11% vs. 77±8%, p=NS). After a Fontan operation in both group 2 and 3, the wall thickness was appropriate for the ventricular size. In group 2, there was a significant decrease in wall thickness during the study (−20±22%, p<0.02), whereas in group 3, there was no significant change.

The patients studied during the perioperative period after a Fontan operation (group 4) showed a major increase in wall thickness 10 days after their operation, especially when related to diastolic dimension (90±9% vs. 175±30%, p<0.001). This inappropriate degree of wall thickness for heart size had decreased significantly by 2 months but was still elevated above normal (126±13%, p<0.001). In the early postoperative measurements, increases in wall thickness correlated closely with decreases in dimension (r=0.88, p<0.005).

Contractility-Stress Data

In the patients who remained palliated (group 1), FS, which is load dependent, increased from 29.7±4.6% to 31.2±4.8% (p<0.05), but VCFc did not change (Figures 3–5). ESSM remained above normal throughout but decreased (67.5±23.3 to 57.7±20.4 g/cm², p<0.05), mainly due to an increase in end-systolic wall thickness (mean increase, 1.6 mm; p<0.02). The load-independent relation of VCFc to ESSM showed no significant change perpendicular to its regression line,[13] suggesting no alteration of left ventricular contractility during the 3-year period.

In patients who had received a Fontan repair during the study period (group 2), FS did not change significantly (30.7±4.8% vs. 32.5±8.5%, p=NS), but

TABLE 3. Patients With Fontan Operation Before 1985 (Group 3)

Patient	Age in 1988 (yr)	Previous surgery age (yr)	Age at Fontan (yr)	Type of connection	BSA 1985 (m^2)	BSA 1988 (m^2)	Ded 1985 mm	Ded 1985 %BSA	Ded 1988 mm	Ded 1988 %BSA	hed 1985 mm	hed 1985 %BSA	hed 1985 %Ded	hed 1988 mm
1	7.6	BT 0.01	2.7	RA-PA(H)	0.670	0.871	43.5	122	42.9	116	9.3	174	127	8.5
2	8.6	–	4.3	RA-RV(H)	0.767	0.954	39.3	108	43.4	115	8.1	143	122	9.0
3	9.2	BT 0.6	5.4	RA-PA(D)	0.745	1.021	39.5	109	41.0	108	8.2	147	123	7.3
4	10.2	BT 0.1	6.3	RA-PA(H)	0.803	0.980	50.2	136	50.2	133	7.0	121	83	8.4
5	11.3	BT 3.5	7.5	RA-PA(D)	0.920	1.232	49.1	131	48.6	125	7.8	127	94	8.6
6	11.4	Band 0.3	3.9	RA-RV(H)	0.943	1.158	41.5	111	49.2	127	6.4	103	91	8.6
7	11.6	–	4.2	RA-RV(H)	0.943	1.153	40.5	108	40.2	104	7.0	113	102	7.1
8	12.0	BT 0.01	4.2	RA-PA(H)	1.060	1.302	64.0	168	52.6	134	5.6	86	121	8.6
9	14.2	BT 0.1	5.8	RA-PA(H)	1.039	1.340	38.2	102	39.2	100	6.0	93	92	8.7
10	14.7	BT 0.3	8.2	RA-RV(H)	1.300	1.632	48.0	122	48.9	121	8.8	124	109	7.6
11	16.2	WAT 1.9	7.0	RA-PA(H)	1.278	1.638	47.9	122	51.1	126	5.9	83	73	9.2
12	16.6	Band 0.2, BT 3.8	11.5	RA-RV(H)	1.342	1.357	47.3	120	43.2	109	7.1	99	89	8.1
13	17.1	BT 0.4, BT 6.0	9.9	RA-RV(X)	1.401	1.612	43.6	110	43.6	108	7.2	98	98	9.6
14	24.9	Brock 2.0, BT 6.8	12.7	RA-PA(C)	1.460	1.420	53.0	133	51.6	130	7.6	102	85	7.1
Mean	13.3		6.7		1.048	1.262	46.1	121	46.1	118	7.3	115	101	8.31
±SD	4.6		3.0		0.265	0.256	6.9	17	4.6	11	1.1	26	17	0.8
p						<0.001			NS (W)	NS (W)				<0.05

Band, pulmonary artery band; BSA, body surface area; %BSA, % of normal for BSA; BT, Blalock-Taussig shunt (classic or modified); Ded, ventricular end-diastolic dimension; %Ded, % of normal for Ded; G, glycosides; hed, ventricular end-diastolic posterior wall thickness; NYHA, New York Heart Association; PA, pulmonary artery; WAT, Waterston shunt; C, Hancock conduit; D, direct anastomosis; H, valved homograft; W, Wilcoxon rank-sum test; X, valved xenograft.

All *p* values refer to a paired *t* test unless stated otherwise.

VCFc increased from 0.929±0.149/sec to 1.050±0.230/sec ($p<0.05$). The relation of VCFc to ESSM showed a significant shift toward enhanced contractility after the Fontan procedure ($p<0.05$).

In the post-Fontan group (group 3 patients), FS and VCFc increased significantly (29.7±4.9% to 34.0±3.4%, $p<0.005$; 1.017±0.173/sec to 1.140±0.131/sec, $p<0.005$). There was a net improvement of

TABLE 4. Patients with Fontan Operation In 1987–1988 (Group 4)

Patient	Age in 1988 (yr)	Previous surgery age (yr)	BSA (m^2)	Ded preop mm	Ded preop %BSA	Ded 10 days mm	Ded 10 days %BSA	Ded 60 days mm	Ded 60 days %BSA	hed preop mm	hed preop %BSA	hed preop %Ded	hed 10 days mm	hed 10 days %BSA	hed 10 days %Ded
1	1.6	BT 0.01	0.463	37.8	113	29.0	87	30.2	90	5.4	118	85	8.2	180	168
2	1.9	BT 0.01	0.531	44.4	129	29.5	86	28.7	83	6.5	134	87	10.9	226	219
3	2.6	BT 0.2	0.550	38.4	111	29.3	85	33.9	99	5.4	110	83	8.5	173	172
4	3.0	–	0.685	44.4	124	34.9	98	33.6	95	7.3	135	97	9.7	180	165
5	4.7	BT 1.5	0.624	51.3	146	41.2	117	36.6	104	8.6	166	99	11.3	218	163
6	6.8	BT 0.01, BT 4.5	0.837	54.2	147	37.6	102	42.4	115	7.0	119	76	13.8	235	218
7	7.9	–	0.928	51.9	139	43.3	116	–	–	7.8	127	89	9.6	156	131
8	12.8	Glenn 5.0, BT 10.2	1.433	53.8	135	44.0	110	46.0	115	9.1	123	101	12.0	162	162
Mean	5.1		0.756	47.0	130	36.1	100	35.9	101	7.1	129	90	10.5	191	175
±SD	3.8		0.315	6.7	14	6.4	13	6.3	12	1.4	17	9	1.9	31	30
p						<0.001	<0.001	<0.001	<0.001				<0.001	<0.001	<0.001

BSA, body surface area; %BSA, % of normal for BSA; BT, Blalock-Taussig shunt (classic or modified); Ded, ventricular end-diastolic dimension; %Ded, % of normal for Ded; G, glycosides; hed, ventricular end-diastolic posterior wall thickness; D, diuretics; FS, fractional shortening; V, vasodilator.

All *p* values refer to a paired *t* test unless stated otherwise.

TABLE 3. *(Continued)*

hed 1988		VCFc (cycles/sec)		ESSM (g/cm²)		Medi-cation	NYHA class
%BSA	%Ded	1985	1988	1985	1988	1988	1988
142	117	0.719	1.166	63.7	32.5	–	I
145	123	1.005	1.236	39.1	31.1	–	I
114	105	0.771	1.062	54.2	50.2	–	I
134	99	1.004	1.060	69.9	66.9	–	I
124	105	1.108	1.182	57.0	49.0	–	I
127	104	1.247	1.154	44.3	51.5	–	I
105	105	1.352	1.460	42.4	41.6	–	I
92	97	0.963	1.031	97.0	63.2	–	I
121	132	0.950	1.093	41.1	36.1	–	II
97	92	1.192	1.290	41.3	35.4	G	II
117	107	0.980	1.102	65.1	50.3	–	I
112	112	0.855	0.914	51.9	63.5	–	II
123	131	1.064	1.100	47.9	37.0	G	I
96	82	1.056	1.161	79.3	79.4	–	I
118	108	1.017	1.140	56.7	49.2		
16	14	0.173	0.131	16.8	15.4		
NS	NS		<0.005 (W)		<0.05		

contractility as analyzed by the VCFc-ESSM relation ($p<0.01$).

There was no discrete pattern and, therefore, no statistical significant change in FS in group 4 patients in the early and late postoperative evaluation (34.6±3.9% vs. 34.5±6.9% [p=NS] vs. 35.7±6.1% [p=NS]). No load-independent data could be obtained in group 4 patients as software required for carotid pulse tracings was not available in the United Kingdom during this period of the study.

TABLE 4. *(Continued)*

hed 60 days			FS preop	FS 10 days	FS 60 days	Medi-cation at	Medi-cation at
mm	%BSA	%Ded	(%)	(%)	(%)	10 days	60 days
6.5	143	128	28.2	27.5	30.0	GDV	G
5.7	118	118	34.1	40.2	40.2	GDV	GD
6.3	128	110	36.1	43.5	33.1	GDV	GD
7.0	130	124	30.8	33.1	39.7	GDV	G
8.8	170	143	35.0	27.3	26.2	GDV	GD
8.0	136	112	41.0	32	42.8	GDV	–
–	–	–	37.1	30.2	–	GDV	–
11	149	142	35.4	42.5	38.1	GDV	GD
7.6	139	126	34.6	34.5	35.7		
1.8	17	13	3.9	6.6	6.1		
<0.01	<0.01	<0.01		NS	NS		

Discussion

Clinicians caring for patients with tricuspid atresia are frequently faced with therapeutic dilemmas. Should patients who are limited by cyanosis and who fulfill criteria for a low-risk Fontan procedure be offered a shunt or a Fontan? Should an elective Fontan procedure be performed in suitable patients who are minimally symptomatic?

Left ventricular hypertrophy and dysfunction are known to compromise long-term survival and symptomatic status in tricuspid atresia as well as to be risk factors for mortality and morbidity after a Fontan operation.[19–22] Several previous studies of ventricular mass and contractility in groups of patients after palliation and after a Fontan procedure have been reported. However, the relevance of the results is uncertain because these studies used indexes of pump function that are themselves influenced by the loading conditions of the heart. In our previous study, we used a load-independent index and demonstrated impairment of contractility in tricuspid atresia before and after a Fontan procedure.[14] From this cross-sectional study, we were unable to determine whether the documented changes were reversible with relief of chronic volume overload and cyanosis. We have now investigated prospectively the effect of the Fontan operation on left ventricular performance before and after surgery in the same patients. These results have been related to changes in size and function seen in patients remaining palliated and in patients retaining their Fontan status during the same period. Our results indicate that a successful Fontan repair results in rapid reduction of left ventricular size, which is followed by regression of hypertrophy so that a normal thickness-to-size ratio can be achieved. Operating at these more favorable dimensions and loading conditions results in early increase in contractility, which improves further in medium-term follow-up.

Dimensions

Increase in preload leads to enhanced ventricular performance and ventricular dilatation in a variety of clinical and experimental situations.[23,24] However, it is well known that despite such adaptation, left ventricular failure develops when volume load is sufficiently severe and prolonged.[25–28] This is precisely the situation imposed on the heart by an arterial systemic-to-pulmonary shunt of sufficient size to provide acceptable relief of cyanosis in patients with tricuspid atresia. A resting arterial oxygen saturation of 85% requires a pulmonary to systemic flow ratio of more than 1.5:1 and, therefore, necessitates a minimum volume load to the left ventricle of approximately 250% of normal.[3,29]

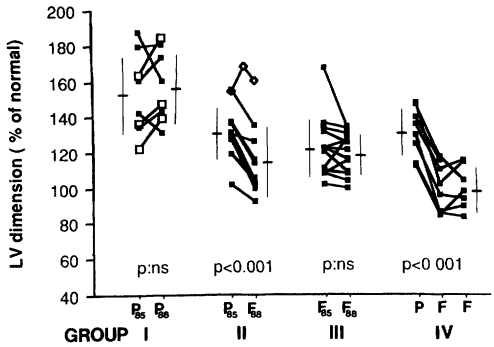

FIGURE 1. *Plots of changes in left ventricular (LV) end-diastolic dimensions expressed as a percentage of normal for body surface area. F, Fontan; P, palliated; 85 and 88 refer to the year of measurement. Open squares in group 1 are patients who received an additional shunt during the study period. The open diamond in group 2 is the patient with atrial flutter (see text).*

Palliated patients in our own and previous studies have greatly increased left ventricular end-diastolic dimensions when compared with normal. During the 3-year follow-up in this study, there was a further increase in dimension in six of eight patients. Progressive dilatation is a major feature of patients left palliated for a long time. Dick and associates[19] reported the clinical course of 101 patients with tricuspid atresia as a gradual attrition due to congestive heart failure that developed in the middle of the second decade, with only very few survivors beyond the third decade.

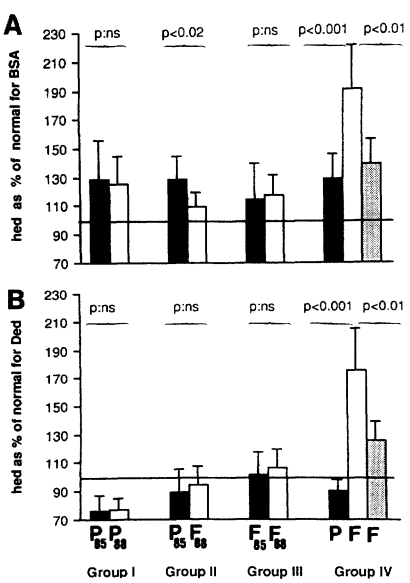

FIGURE 2. Panel A: *Bar graph of left ventricular posterior wall thickness (hed) expressed as a percentage of that predicted by body surface area (BSA).* Panel B: *Bar graph of hed expressed as a percentage of that predicted by ventricular end-diastolic dimension (Ded). F, Fontan; P, palliated; 85 and 88 refer to the year of measurement.*

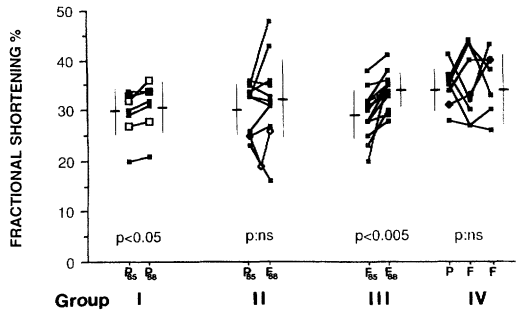

FIGURE 3. *Plots of changes in fractional shortening. F, Fontan; P, palliated; 85 and 88 refer to the year of measurement. [Open squares in group 1 are patients who received an additional shunt during the study period. The open diamond in group 2 is the patient with atrial flutter (see text).]*

Our results confirm those of Nakae et al,[12] who reported a significant decrease of the left ventricular end-diastolic dimension in six patients after a Fontan procedure. Similar observations have been made after complete relief of left ventricular volume overload in adults with aortic regurgitation or mitral regurgitation, in children after ventricular septal defect closure, and in experimental conditions with aortocaval fistulae.[30-33] In our patients, left ventricular dimensions rapidly decreased after Fontan repair to reach a new steady state in most by 10 days. Studies in the post-Fontan group show that this improvement is maintained over a longer period provided that no adverse factors, such as arrhythmia, are present.

Interestingly, the early reduction in size after the Fontan procedure was insufficient to reach levels normal for body surface area. However, with time, values showed a tendency to normalize as the patient's body surface area increased. When evaluated at least 6 months after repair as in groups 2 and 3, 10 of 25 patients had left ventricular dimensions that were within normal range for their body surface area. Of special interest are the patients of group 4 because they are more representative for current medical and surgical management (less palliative procedures, Fontan operation at younger age): in the early postoper-

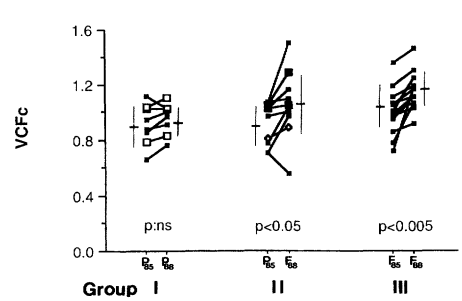

FIGURE 4. *Plots of changes in VCFc. F, Fontan; P, palliated; 85 and 88 refer to the year of measurement. Open squares in group 1 are patients who received an additional shunt during the study period. The open diamond in group 2 is the patient with atrial flutter (see text).*

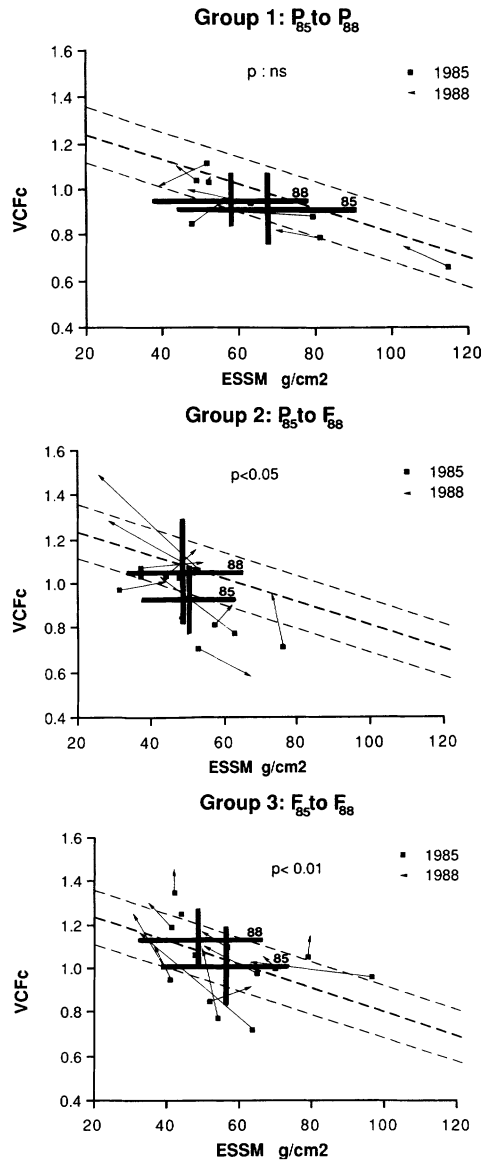

FIGURE 5. *Plots of endocardial rate-corrected circumferential fiber shortening (VCFc) as a function of meridional end-systolic stress (ESSM) for groups 1, 2, and 3. Normal values ±2 SD from Colan et al.[13] Changes parallel to the mean regression line reflect altered loading conditions; changes perpendicular to the mean regression line reflect altered contractility.*

ative period, six of eight had ventricles with normal end-diastolic dimension.

The palliated patients in our study showed myocardial hypertrophy that was not inappropriately high for the cavity size of the ventricle, a situation comparable to that observed in other situations with chronic volume overload.[30] Also, when the preload stress was removed by the Fontan operation, the ventricular thickness gradually decreased to values appropriate for the reduced dimensions.

The rate of decrease in cavity size and wall thickness after the Fontan operation were very different, with reduction of thickness taking much longer than reduction in size. As a result, the ventricles were inappropriately thick for their dimensions in the early postoperative period. This may have implications for the diastolic properties of the left ventricle in the acute postoperative period and requires further study.

Contractility

The major changes in dimension and thickness of the left ventricle that follow a Fontan procedure make evaluation of ventricular systolic function with load-dependent indices difficult. When ventricular dimension and wall stress are increased, ejection fraction (EF) and FS underestimate contractility.[13] Furthermore, when dimensions alter during the study period, EF and FS may fail to reflect major changes in contractility.

Sanders et al investigated 11 patients before and after a Fontan procedure with angiocardiography.[7] The EF remained normal in 10 patients and decreased in one, with no overall change for the group. Using radionuclide angiography, Hurwitz et al showed no significant change in mean EF during the early postoperative period; however, at restudy 2 years after surgery, the EF had increased significantly to be within the normal range in all six patients reported.[34] Our own results using a load-dependent index confirm these findings with no significant change of the FS in the early postoperative phase after a Fontan procedure. However, with the gross changes in loading conditions, it would be inappropriate to conclude there was no change in contractility. We, therefore, also made comparisons of the relation of VCFc to ESSM as Colan et al has advocated to produce an index relatively independent of loading conditions but sensitive to altered inotropic state.[13] This relation revealed that there was a significant improvement in contractility by 6 months after a Fontan procedure. It is encouraging to note that the contractility improved further in the medium-term follow-up as has also been shown for patients with aortic regurgitation and a moderate degree of preoperative left ventricular dysfunction.[30,35] No improvement occurred in the single patient who survived a Fontan repair but had profoundly decreased contractility at initial evaluation. Despite medical therapy, she died in congestive heart failure, suggesting that as in aortic regurgitation, there may be a "point of no return." We could not, however, evaluate this systematically because the selection criteria for the Fontan operation precluded surgery in patients with left ventricular dysfunction.

Limitations of the Study

The limitations of the VCFc-ESSM method have previously been discussed in detail.[36] The assumptions of the model include that the short-axis dimensions are representative for ventricular size and global left ventricular performance. As the ventricle in tricuspid atresia is more spherical than normal,[3,37] this is likely to be the case. All patients were screened for regional wall motion abnormalities to accept the

M-mode echocardiogram as representative for the global ventricular function. At initial evaluation in 1985, five patients of the post-Fontan group had mild septal motion abnormalities. We had to exclude one of these patients because the septal dyskinesia in the 1988 evaluation had increased, although overall left ventricular function appeared well preserved; the other four patients showed subsequent improvement and were, therefore, included. No other regional dyskinesia was observed in the other groups.

The ESSM model assumes the ventricular chamber to be thin walled. Although all patients had some degree of hypertrophy when the ventricular thickness was related to their BSA, the ventricular wall appeared to be appropriate or even too thin when related to the ventricular size.

We could not determine the relation between preoperative ventricular function and long-term improvement after the Fontan operation over a wide spectrum of left ventricular function in patients with tricuspid atresia because poor ventricular function precluded acceptance for Fontan. The data from this study relate to a relatively small number of patients selected by clinical practice and should not be extrapolated to all patients with tricuspid atresia.

Four patients died during the study interval, two at the time of the Fontan operation. This serves to remind clinicians that the Fontan procedure can still not be performed with the predictably low risk of a systemic-to-pulmonary shunt. Interestingly, there was a relation between age and decreased contractile function in palliated patients; we could not attempt to relate age at surgery to change in postoperative ventricular function.

Conclusion

This prospective study has examined the effect of palliation and Fontan operation on ventricular size and function in patients with tricuspid atresia. Abnormal size and contractile function is present in most patients with palliation alone. A successful Fontan operation can significantly improve these abnormalities and ventricular contractility may continue to improve, at least in the medium term. Because left ventricular dysfunction is the most important determinant of long-term survival, these findings suggest that a Fontan operation may provide a beneficial long-term investment and argue in favor of such a repair, even in mildly symptomatic patients who fulfill the requirements for a low-risk operation.

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KEY WORDS • Fontan procedure • contractility • volume loading