SURGERY FOR CONGENITAL HEART DISEASE

LATE RESULTS OF SYSTEMIC ATRIOVENTRICULAR VALVE REPLACEMENT IN CORRECTED TRANSPOSITION

From December 1964 to October 1993, 40 patients (aged 5 months to 70 years, mean 21.8 years, median 13.6 years) with corrected transposition and systemic atrioventricular valve insufficiency underwent replacement ($n = 39$) or repair ($n = 1$) of the systemic atrioventricular valve. Thirty-nine patients had situs solitus and 1 had situs inversus. Associated anomalies included Ebstein's malformation of the systemic atrioventricular valve ($n = 22$), ventricular septal defect ($n = 19$), and pulmonary stenosis ($n = 14$). Preoperatively, 16 patients (40.0%) had complete heart block and 27 patients (67.5%) were in New York Heart Association functional classes III and IV. The early mortality was 10.0% ($n = 4$) and 8 patients died subsequently. The principal cause of death in all 12 patients was systemic ventricular failure. Overall survival including early mortality was 78.0% at 5 years and 60.7% at 10 years; survival excluding early mortality was 86.7% at 5 years and 67.5% at 10 years. Survivorship correlated with preoperative systemic ventricular ejection fraction of 44% or more ($p < 0.001$) and later interval of operation (9 deaths in 15 patients before 1981 versus 3 deaths in 25 patients subsequently) ($p = 0.06$). There were no cases of surgically induced complete heart block. Two patients underwent late reoperations related to the systemic atrioventricular valve prosthesis. Follow-up extended to 26.0 years (median 4.7 years). At last follow-up, 18 of the 28 survivors were in New York Heart Association functional class I, 9 were in class II, and 1 was in class III. We conclude that the results of systemic atrioventricular valve replacement in corrected transposition have improved significantly during the past decade. To preserve systemic ventricular function, we suggest operation be considered at the earliest sign of progressive ventricular dysfunction as assessed by serial clinical evaluation and echocardiography. (*J THORAC CARDIOVASC SURG* 1995;109:642-53)

Jacques A. M. van Son, MD* (by invitation), Gordon K. Danielson, MD, James C. Huhta, MD,** Carole A. Warnes, MD (by invitation), William D. Edwards, MD (by invitation), Hartzell V. Schaff, MD, Francisco J. Puga, MD, and Duane M. Ilstrup, MS (by invitation), *Rochester, Minn.*

From the Division of Thoracic and Cardiovascular Surgery, Division of Cardiovascular Diseases, Division of Anatomic Pathology, and Department of Medical Statistics and Epidemiology, Mayo Clinic, Rochester, Minn.

Read at the Seventy-fourth Annual Meeting of The American Association for Thoracic Surgery, New York, N.Y., April 24-27, 1994.

Address for reprints: G. K. Danielson, MD, Division of Thoracic and Cardiovascular Surgery, Mayo Clinic, Rochester, Minnesota 55905.

*Current address: Division of Cardiovascular Surgery, UCSF, San Francisco, CA 94143.

**Current address: Perinatal Cardiology, Pennsylvania Hospital, Philadelphia, PA 19107.

Copyright © 1995 by Mosby-Year Book, Inc.

0022-5223/95 \$3.00 + 0 12/6/61886

Congenitally corrected transposition of the great arteries (biventricular heart with atrioventricular [AV] discordance and ventriculoarterial discordance) has been reported to account for 1.4% of congenital heart disease.¹ Corrected transposition may occur in situs solitus (Van Praagh {S,L,L}) or in situs inversus (Van Praagh {I,D,D}). Abnormalities of the left-sided (in situs solitus) or right-sided (in situs inversus) morphologically tricuspid systemic AV valve in corrected transposition have been described in pathology²⁻⁶ and clinical reports.⁷⁻²⁰ The spectrum of valvular abnormalities ranges from minimal deformity such as inferior displacement of the coaptation point to severe dysplasia of one or

Table I. Associated cardiovascular anomalies (29 patients)

Anomaly	Number	Percent
Ventricular septal defect	19	47.5
Atrial septal defect	16	40.0
Pulmonary stenosis	14	35.0
Pulmonary AV valve insufficiency	4	10.0
Coarctation of aorta	3	7.5
Aortic stenosis	2	5.0
Aneurysm of ventricular septum	2	5.0
Patent ductus arteriosus	1	2.5
Criss-cross AV relationships	1	2.5
Situs inversus totalis	1	2.5
Double-orifice systemic AV valve	1	2.5
Unroofed coronary sinus	1	2.5
Persistent left superior vena cava	1	2.5
Hypoplasia of right pulmonary artery	1	2.5
Stenosis of left pulmonary artery	1	2.5
Aberrant LAD from RCA	1	2.5
Accessory conduction pathway	1	2.5

AV, Atrioventricular; LAD, left anterior descending coronary artery; RCA, right coronary artery.

more leaflets, and valvular incompetence ranges from none to severe. Apical displacement of the septal leaflet and other abnormalities of "left-sided" Ebstein's malformation are common.⁶ In a previous clinical study, we¹⁹ reported a 33% incidence of systemic AV valve insufficiency in corrected transposition; this lesion has a negative effect on survival whether occurring alone or in association with other defects. The relative importance of systemic AV valve insufficiency versus systemic (morphologically right) ventricular dysfunction, which is often found in association, is not clear. To learn more of this relationship and the outcome of surgical patients, we reviewed the late results of all the patients with corrected transposition who had systemic AV valve repair or replacement at our institution.

Patients and methods

Between December 1964 and October 1993, 40 patients with corrected transposition and systemic AV valve insufficiency had repair or replacement of the systemic AV valve at the Mayo Clinic. Patients with AV discordance associated with double-outlet right ventricle were excluded. Information from the records of these patients was collected and analyzed by means of a computer database that included sex; date of birth; associated cardiovascular anomalies; previous and concomitant cardiovascular procedures; cardiac rhythm, symptoms, and activity level before the operation and at follow-up; and preoperative and postoperative catheterization and echocardiography data, especially grade of preoperative systemic AV valve insufficiency and ejection fraction of the systemic ventricle. Additional follow-up data were col-

Table II. Previous cardiovascular procedures (16 patients)

Procedure	Number	Percent
Pacemaker implantation	8	20.0
Closure ventricular septal defect	7	17.5
Closure atrial septal defect*	5	12.5
Pulmonary ventricle-pulmonary artery conduit	3	7.5
Modified Blalock-Taussig shunt	3	7.5
Coarctation repair	3	7.5
Pulmonary artery banding	3	7.5
Pulmonary artery debanding	1	2.5
Systemic AV valve annuloplasty	1	2.5
Closure patent ductus arteriosus	1	2.5
Pulmonary valvotomy	1	2.5
Cardiac exploration	1	2.5

*Closure was performed surgically in 4 patients and with a clamshell device in 1 patient.

lected by examination and questionnaires completed by patients/parents and referring physicians. Telephone calls were made to complete data acquisition when necessary.

Twenty-five patients (62.5%) were male and 15 (37.5%) were female. The ages of the patients at operation ranged from 5 months to 70 years (mean 21.8 ± 19.5 years) (median 13.6 years). Thirty-nine patients had situs solitus with a left-sided morphologically tricuspid systemic AV valve and one patient had situs inversus totalis (situs inversus with mirror-image dextrocardia) and a right-sided morphologically tricuspid systemic AV valve. Not all of the valves were analyzed before excision; however, 22 (55.0%) were clearly seen to have "left-sided" Ebstein's malformation of the systemic AV valve. Twenty-nine patients (72.5%) had one or more associated cardiovascular anomalies (Table I). Sixteen patients had one or more previous cardiovascular procedures elsewhere or at our institution (Table II).

Two patients (5.0%) had no symptoms; 11 (27.5%) were in New York Heart Association (NYHA) functional class II, 21 (52.5%) were in class III, and 6 (15.0%) were in class IV. One patient in NYHA class II had bacterial endocarditis 3 months before the operation. Findings in the 27 patients in NYHA classes III and IV included mild to severe peripheral edema ($n = 11$), renal failure (serum creatinine level > 2 mg/dl) ($n = 6$), cyanosis ($n = 2$), transient ischemic attacks associated with atrial fibrillation ($n = 2$), and liver failure (total bilirubin level > 3 mg/dl) ($n = 2$).

Normal sinus rhythm was present in 24 patients (60.0%); 16 (40.0%) had complete AV block. Heart block was present at birth in 3 patients (7.5%), developed spontaneously in 11 patients (27.5%), and followed a previous cardiac procedure elsewhere in 2 patients (5.0%). Electrocardiographic abnormalities are summarized in Table III.

All patients except for 2 recent ones had preoperative cardiac catheterization and angiography (the 2 exceptions had echocardiography only). Twenty patients had preoperative echocardiography (Fig. 1). Systemic AV valve insufficiency was graded angiographically and/or echocar-

Table III. *Electrocardiographic abnormalities*

Abnormality	Number	Percent
Complete AV block	16	40.0
First-degree AV block	13	32.5
Atrial fibrillation	6	15.0
Right bundle branch block	3	7.5
Left bundle branch block	2	5.0
Atrial flutter	1	2.5
Second degree AV block	1	2.5
Ventricular preexcitation (WPW)	1	2.5

WPW, Wolff-Parkinson-White.

diographically as trivial, mild, moderate, or severe. By either or both techniques, 36 patients had severe and 4 had moderate systemic AV valve insufficiency and were considered surgical candidates. The preoperative ejection fraction of the systemic ventricle ranged from 20% to 62% (mean 47.9%, median 50%). Thirty-six patients were studied by echocardiography after the operation.

Early deaths were defined as those occurring within 30 days of operation or during the surgical hospitalization, whichever was longer. Survival estimates were made with the Kaplan-Meier product limit method, and comparisons between survival distributions were made with the log rank test. The effects of continuous variables, such as ejection fraction, on survival were evaluated with Cox proportional hazards models. Differences were considered significant when $p \leq 0.05$.

Operative techniques. Standard techniques for cardiopulmonary bypass were used; cold potassium or blood cardioplegic solution has been used for myocardial protection since 1977. In 39 patients with situs solitus, replacement or repair of the systemic AV valve was accomplished through a standard left atriotomy anterior to the right pulmonary veins in 38 and through a right atriotomy and transeptal approach in 1. In the 1 patient with situs inversus totalis, the insufficient right-sided systemic AV valve was approached transeptally through the left-sided morphologically right atrium. Prosthetic valves used in the 39 patients in whom the systemic AV valve was replaced included Starr-Edwards (Baxter Healthcare Corp., Edwards Division, Irvine, Calif.) ($n = 24$), St. Jude Medical (St. Jude Medical, Inc., St. Paul, Minn.) ($n = 8$), Carpentier-Edwards (Baxter) ($n = 4$), and Hancock (Johnson & Johnson Cardiovascular, King of Prussia, Pa.) ($n = 3$). One patient with moderate systemic AV valve insufficiency underwent an annuloplasty of the valve. Twenty-nine patients underwent one or more concomitant cardiovascular procedures (Table IV).

Results

Early results. There were 4 early deaths (10.0%); 1 patient died during the operation and 3 died on the first to the fifth postoperative day. In all instances, the cause of death was failure of the systemic ventricle. Postoperative morbidity included transient arrhythmia ($n = 11$), reoperation for bleeding ($n = 3$), and wound infection ($n = 1$).

Surgically induced complete heart block did not develop.

Late results. Follow-up extended to 26.0 years (median 4.7 years) and was complete in 37 patients (92.5%); 3 overseas patients were lost to follow-up after 5, 6, and 12 years, respectively. Eight patients (20.0%) died late of congestive heart failure (Table V). Survival including early mortality was 78.0% at 5 years and 60.7% at 10 years, respectively (Fig. 2). Survival excluding early mortality was 86.7% at 5 years and 67.5% at 10 years, respectively.

The preoperative systemic ventricular ejection fraction correlated closely with survival. All 12 patients who died had a systemic ventricular ejection fraction of less than 44% (range 25% to 42%, mean 34.6%), whereas for survivors the range was 20% to 62%, mean 47.9% ($p < 0.001$). Eleven of the 12 nonsurvivors (92%) died within 8.4 years of operation. The 5-year and 10-year survivals in the patients with an ejection fraction of less than 44% were 48.9% and 19.5%, respectively (Fig. 3). In contrast, the 5-year and 10-year survivals for patients with an ejection fraction of 44% or more were 100% ($p < 0.001$).

A second factor that correlated with survival was interval of operation. Of the 15 patients who underwent operation before 1981, 9 (60.0%) died (3 early and 6 late), versus 3 (1 early and 2 late) of the 25 patients (12.0%) who underwent operation subsequently; this difference approached statistical significance at $p = 0.06$. Patients who underwent operation before 1981 versus those who underwent operation subsequently differed with regard to ejection fraction (median 40% versus 50%, respectively; $p = 0.04$), age at operation (median 13.2 years versus 21.6 years, respectively; $p = 0.39$), and presence of preoperative complete heart block (5 of 15 patients versus 11 of 25 patients; $p = 0.74$). Preoperative NYHA functional classes I and II versus classes III and IV correlated less closely with survival than interval of operation ($p = 0.09$) (Fig. 4).

Preoperative variables that did not appear to influence survival were previous operation ($p = 0.13$), pacemaker dependency ($p = 0.26$), complete heart block ($p = 0.31$), sinus rhythm ($p = 0.42$), associated ventricular septal defect ($p = 0.45$), and associated pulmonary stenosis ($p = 0.47$).

One patient underwent two replacements of porcine systemic AV valve prostheses and reclosure of a ventricular septal defect 2 and 5 years, respectively, after primary valve replacement at age 5 years. Four other patients underwent one reopera-

Fig. 1. **A**, Echocardiogram in the apical four-chamber view in a patient with corrected transposition and situs solitus. The morphologically right ventricle (*mRV*) is identified partly by its heavy trabeculations (*small arrows*) and also by the tricuspid valve, which is the more apically placed of the two AV valves (*large arrows*). This arrangement of the cardiac crux is the mirror-image of normal. The *arrowheads* point to the pulmonary veins. *AS*, Atrial septum; *RA*, right atrium; *LA*, left atrium; *mLV*, morphologically left ventricle; *VS*, ventricular septum (From Seward JB, Tajik AJ, Edwards WD, et al. Two-Dimensional Echocardiographic Atlas, Vol. 1. Congenital Heart Disease. New York, Springer-Verlag, 1987. By permission, Mayo Foundation.) **B**, Specimen of corrected transposition cut in the same plane. There is AV discordance. Note the left-sided tricuspid AV valve that inserts lower than the right-sided mitral valve. (From Hagler DJ, Tajik AJ, Seward JB, Edwards WD, Mair DD, Ritter DG. Atrioventricular and Ventriculoarterial Discordance [Corrected Transposition of the Great Arteries]: Wide Angle Two-Dimensional Echocardiographic Assessment of Ventricular Morphology. Mayo Clin Proc 56:591-600, 1981. By permission, Mayo Foundation.) **C**, An echocardiogram from the apical four-chamber view demonstrating Ebstein-like abnormality of the left AV (tricuspid) valve in a patient with corrected transposition. Note the marked inferior displacement of the valve (*arrow*). Abbreviations are the same as for **A**. (From Seward JB, Tajik AJ, Edwards WD, et al: Two-Dimensional Echocardiographic Atlas, Vol. 1. Congenital Heart Disease. New York, Springer-Verlag, 1987. By permission, Mayo Foundation.)

**Table IV. Concomitant cardiovascular procedures
(29 patients)**

Procedure	Number	Percent
Closure ventricular septal defect	12	30.0
Closure atrial septal defect	11	27.5
Pacemaker implantation	6	15.0
Electrophysiologic mapping	3	7.5
Pulmonary AV valve annuloplasty	3	7.5
Closure modified Blalock-Taussig shunt	3	7.5
Pulmonary valvotomy	2	5.0
Pulmonary ventricle-pulmonary artery conduit	2	5.0
Pulmonary valve replacement	2	5.0
Resection subpulmonary stenosis	2	5.0
Aortic valve replacement	2	5.0
Repair aneurysm of ventricular septum	2	5.0
Pulmonary artery debanding	2	5.0
Closure residual atrial septal defect	1	2.5
Cryoablation accessory pathway	1	2.5
Ligation left superior vena cava	1	2.5
Correction unroofed coronary sinus	1	2.5
Patch repair stenosis of left pulmonary artery	1	2.5

tion each for reasons other than replacement of a systemic AV valve prosthesis (Table VI).

Complete heart block did not develop in any of the 15 survivors who did not have complete heart block before the operation. Important ventricular arrhythmias were present in 5 patients who died late and in 1 patient who received an automatic internal cardioverter defibrillator. At latest follow-up in the 28 long-term survivors (range 1 month to 26 years, mean 6.3 years, median 4.7 years), 8 had improved two functional classes, 17 had improved one functional class, and 3 remained in the same functional class. The latest NYHA functional class in the 28 long-term survivors was class I in 18, class II in 9, and class III in 1. The 1 patient who underwent valvuloplasty of the systemic AV valve is in NYHA class I with only mild systemic AV valve insufficiency at 16 years' follow-up.

In the 36 patients who had early or late postoperative echocardiographic follow-up, the mean systemic ventricular ejection fraction was 41.6% (range 15% to 60%), whereas the mean preoperative ejection fraction in the same patients was 45.8% (range 20% to 62%) ($p < 0.001$, paired t test). At latest echocardiographic follow-up, the 1 patient in NYHA class III had an ejection fraction of 15%, compared with a preoperative value of 20%; this patient currently is a candidate for heart transplantation.

Table V. Late deaths

Patient	Age at death (yr)	Postop interval (yr)	Cause	Autopsy
1	37	7	CHF, CM, ventricular arrhythmia, pulmonary AV valve insufficiency	Yes
2	21	8	CHF, CM, heart transplantation	Yes
3	55	4 $\frac{1}{2}$	CHF, ventricular arrhythmia	Yes
4	36	26	CHF, CM, ventricular arrhythmia	Yes
5	6	4	CHF, pannus ingrowth systemic AV valve prosthesis	Yes
6	5	3 $\frac{1}{2}$	CHF, ventricular arrhythmia	Yes
7	20	2 $\frac{1}{2}$	CHF, CM, ventricular arrhythmia, thrombosis systemic AV valve prosthesis	Yes
8	25	7	CHF	No

CHF, Congestive heart failure; CM, cardiomyopathy.

Discussion

Developmental abnormalities of the systemic AV valve in corrected transposition are so common that they may be considered an intrinsic part of the pathophysiology of this condition. The incidence of valve insufficiency has been reported to be approximately 33% in our experience¹⁹ and that of others.^{3, 13-16, 20} A "left-sided" Ebstein-like anomaly is the most frequently observed malformation of the valve^{5, 6} (Fig. 1, C and Fig. 5). However, significant differences exist between left-sided Ebstein's anomaly and the classic right-sided variety: (1) the AV anulus is not dilated, (2) the anterior leaflet is not enlarged and may be cleft (rather than saillike), (3) the morphologic right ventricle is either small or normal-sized and does not begin to dilate until late in the clinical course, and (4) the atrialized right ventricle is not appreciably thinned.^{6, 21, 22} Similarities between the two varieties include apical displacement of the septal and posterior leaflets, leaflet dysplasia with varying degrees of failure to delaminate from the ventricle, and abnormal papillary muscles and short chordae.^{5, 6, 22, 23} These abnormalities create potential or actual AV valve regurgitation.

It is of interest that severe systemic AV valve insufficiency has developed in patients with corrected transposition after repair of ventricular septal defect or other anomalies, both in our experience and in that of others.^{9, 11, 13, 14} This is thought to be due to some ventricular dilatation, which results from increased afterload or transiently decreased

Fig. 2. Survivorship with and without early mortality in patients with corrected transposition having replacement ($n = 39$) or repair ($n = 1$) of the systemic AV valve. Expected survival in an age- and sex-matched control population is also shown.

Fig. 3. Overall survivorship of patients with preoperative systemic ventricular ejection fraction (EF) of 44% or more versus those with an ejection fraction of less than 44%.

ventricular function after cardiopulmonary bypass, causing the marginally compensated systemic AV valve to become incompetent. Systemic AV valve insufficiency has also been observed after spontaneously or iatrogenically induced complete heart block.^{9, 13, 14} In these patients, a slower heart rate may produce an increased stroke volume and enough ventricular dilatation to render the marginally compensated systemic AV valve incompetent. The development of systemic AV valve insufficiency

is undoubtedly multifactorial, inasmuch as 60% of the patients in our series did not have a previous cardiac operation and 60% did not have preoperative complete heart block.

The AV node and bundle of His in corrected transposition with situs solitus differ from the normal. Although a regular (posterior) AV node may be present in front of the coronary sinus ostium at the apex of the triangle of Koch, the penetrating bundle rarely extends from it because of septal

Fig. 4. Overall survivorship of patients in preoperative NYHA classes I and II versus those in classes III and IV.

Table VI. Reoperations

Reoperation	No. of patients
Replacement systemic AV valve prosthesis and reclosure VSD ($\times 2$)	1
Repair perivalvular leak systemic AV valve prosthesis	1
Heart transplant	1
Replacement pulmonary AV valve	1
Insertion automatic internal cardioverter defibrillator	1

VSD, Ventricular septal defect.

malalignment.²⁴ The bundle of His usually arises from a second, more anteriorly and superiorly located AV node and courses anterior to the pulmonary artery and down the morphologically left ventricular side of the septum. When a ventricular septal defect is present, the His bundle passes anterior to the defect. The abnormal origin and course of the conduction system predispose it to anatomic discontinuity and fibrosis with resulting complete heart block.²⁴⁻²⁷

In contrast, in patients with situs inversus with mirror-image dextrocardia, the penetrating bundle nearly always originates from the regular (posterior) AV node and courses on the left ventricular side of the septum (and on the posteroinferior margin of the ventricular septal defect, when present).^{11, 28, 29} The occurrence of spontaneous complete heart block has been observed to be less frequent in corrected transposition with situs inversus (7%)¹⁸

than with situs solitus (22% with increments of 2% per year).³⁰

The incidence of complete heart block after repair of associated cardiac anomalies in corrected transposition has been reported as high as 40%.^{9, 14, 17} Current understanding of the abnormal conduction system in corrected transposition and the development of surgical techniques to avoid injury to the conduction tissue³¹⁻³⁴ have reduced the risk of surgically induced heart block considerably. For isolated systemic AV valve replacement, the risk of heart block should be low, because the anulus is relatively far removed from the conduction system; in the present series, surgically induced complete heart block did not develop in any patient.

Because of the atypical Ebstein's characteristics that are encountered in the systemic AV valve of patients with corrected transposition, valve plication and annuloplasty techniques that can be successfully applied to more than half of patients with classic right-sided Ebstein's anomaly³⁵ are not applicable. Only 1 patient in the current series underwent successful repair of the systemic AV valve, and the valve insufficiency was only moderate. In general, repair techniques are associated with a high rate of failure (up to 75%) and the need for reoperation for valve replacement.^{13, 15, 20} Although a low-profile prosthesis has generally been advocated and seems reasonable on the basis of theoretical grounds, the high-profile Starr-Edwards valve was used successfully in many patients in this series.

Fig. 5. Ebstein's malformation of left-sided systemic AV (morphologically tricuspid) valve in corrected transposition of the great arteries. **A**, Septal (S) and posterior (P) leaflets show mild downward displacement from level of true anulus (*small dashed line*) to level of functional anulus (*large dashed line*). **B**, Cross-sectional slice through atrial (AS) and ventricular (VS) septa demonstrates downward displacement of tricuspid anulus (*arrow*). mLA, Morphologically left atrium; mLV, morphologically left ventricle; mRA, morphologically right atrium; mRV, morphologically right ventricle; VSD, ventricular septal defect.

Patients with corrected transposition unaccompanied by significant associated lesions often have normal systemic ventricular function with an adequate increase in cardiac output during exercise well into late adulthood.³⁶⁻³⁸ However, this is primarily due to an increase in heart rate, and stroke volume is increased to a lesser extent.^{37, 39} Systemic AV valve insufficiency imposes a volume overload on the already existing pressure overload of the morpho-

logically right systemic ventricle. These factors are associated with a tendency for systemic ventricular function to deteriorate progressively, which may occur as early as the first decade of life.^{17, 21, 40} Other possible factors contributing to deterioration of systemic ventricular function are intrinsic abnormalities of the morphologically right ventricle and development of complete heart block.^{17, 19, 30}

Systemic AV valve insufficiency in corrected

transposition causes an underestimation of systemic ventricular dysfunction. Systolic function of the systemic ventricle as determined by ejection phase indices (e.g., ejection fraction) may appear better than it actually is because of the reduced afterload caused by the systemic AV valve insufficiency.^{21, 40}

The reported early mortality for systemic AV valve replacement in corrected transposition has ranged up to 25%.^{*} The results of this study indicate that the early mortality has improved considerably during the past decade. We believe the improved results are due primarily to our change in philosophy toward earlier surgical intervention to preserve systemic ventricular function, as evidenced by a higher median ejection fraction in the patients operated on in the more recent experience. Perhaps improved methods of myocardial preservation with antegrade and retrograde cold blood cardioplegia have also been a factor in the improved results.

Late results are also encouraging. In 25 of 28 surviving patients, the functional status improved one to two classes. However, at early and late follow-up, there was no improvement in the mean ejection fraction compared with the mean preoperative ejection fraction. All survivors who are currently in NYHA class II or III had preexisting moderately or severely depressed systolic systemic ventricular function associated with long-standing systemic AV valve insufficiency. Therefore, we emphasize the importance of timely systemic AV valve replacement in corrected transposition to preserve ventricular function, and we suggest that operation be considered at the earliest signs of progression of symptoms or evidence of progressive systemic ventricular deterioration such as an enlarging ventricle and left atrium, development of pulmonary hypertension, and appearance of atrial arrhythmias. It is anticipated that improved monitoring of ventricular function with serial echocardiographic/Doppler hemodynamic assessment will result in further improvement of the long-term outcome of such patients.

In addition to being affected by the systemic ventricular function and the development of systemic AV valve insufficiency, the long-term fate of these patients is influenced by the effects of associated anomalies and the need for reoperation for replacement of stenotic pulmonary ventricle-pulmonary artery conduits or replacement of malfunctioning prosthetic valves.

Several suggestions have been made to avoid the problems of late deterioration of systemic ventricular function by rerouting intracardiac blood so that the morphologically left ventricle becomes the systemic ventricle. One recommendation applicable to patients with corrected transposition, ventricular septal defect, and pulmonary stenosis or atresia is an atrial switch procedure combined with a "Rastelli-type" operation. In this procedure the ventricular septal defect is closed in a manner that directs blood from the morphologically left (pulmonary) ventricle into the aorta, the origin of the pulmonary trunk is closed, and the morphologically right (systemic) ventricle is connected to the pulmonary trunk with or without a valved conduit.⁴¹⁻⁴³ Others^{44, 45} have suggested, for patients with corrected transposition, ventricular septal defect, and no pulmonary stenosis, that a double switch operation be performed: an atrial switch of the Senning or Mustard type combined with an arterial switch of the Jatene type. Possible problems with these procedures include higher initial operative mortality, late complications of conduits, late complications of atrial switch procedures including pulmonary and systemic venous obstruction, arrhythmias, and reduced ejection fraction of the morphologically left ventricle.^{36, 44} More experience is necessary to evaluate the relative roles of these newer approaches in the surgical management of associated anomalies in corrected transposition.

In conclusion, during the past decade the early and late survival in patients who have undergone systemic AV valve replacement in corrected transposition has improved appreciably. The long-term outcome in corrected transposition is mainly determined by systemic ventricular function. So that this function can be preserved, consideration should be given to replacing the systemic AV valve at the earliest signs of progressive ventricular dysfunction, which is best diagnosed by serial clinical evaluation and echocardiographic/Doppler hemodynamic assessment.

We appreciate the opportunity of including data from patients operated on by Dr. J. W. Kirklin, Dr. D. C. McGoon, and Dr. R. B. Wallace.

REFERENCES

1. Fontana RS, Edwards JE. Frequency of occurrence of various anatomic types of malformations among patients with congenital cardiac disease. In: *Congenital cardiac disease: a review of 357 cases studied pathologically*. Philadelphia: WB Saunders, 1962:28-65.

*References 8, 9, 11, 13, 14, 16, 17, 20, 32.

2. Becu LM, Swan HJC, DuShane JW, Edwards JE. Ebstein malformation of the left atrioventricular valve in corrected transposition of the great vessels with ventricular septal defect. *Proc Staff Meet Mayo Clin* 1955;30:483-90.
3. Schiebler GL, Edwards JE, Burchell HB, DuShane JW, Ongley PA, Wood EH. Congenital corrected transposition of the great vessels: a study of 33 cases. *Pediatrics* 1961;27(Suppl):I851-88.
4. Van Mierop LHS, Alley RD, Kausel HW, Stranahan A. Ebstein's malformation of the left atrioventricular valve in corrected transposition, with subpulmonary stenosis and ventricular septal defect. *Am J Cardiol* 1961;8:270-4.
5. Allwork SP, Bentall HH, Becker AE, et al. Congenitally corrected transposition of the great arteries: morphologic study of 32 cases. *Am J Cardiol* 1976;38:910-23.
6. Anderson KR, Danielson GK, McGoon DC, Lie JT. Ebstein's anomaly of the left-sided tricuspid valve: pathological anatomy of the valvular malformation. *Circulation* 1978;58(Suppl):I87-91.
7. Friedberg DZ, Nadas AS. Clinical profile of patients with congenital corrected transposition of the great arteries: a study of 60 cases. *N Engl J Med* 1970;282:1053-9.
8. Bonfils-Roberts EA, Guller B, McGoon DC, Danielson GK. Corrected transposition: surgical treatment of associated anomalies. *Ann Thorac Surg* 1974;17:200-9.
9. Fox LS, Kirklin JW, Pacifico AD, Waldo AL, Barger LM Jr. Intracardiac repair of cardiac malformations with atrioventricular discordance. *Circulation* 1976;54:123-7.
10. Krongrad E, Ellis K, Steeg CN, Bowman FO Jr, Malm JR, Gersony WM. Subpulmonary obstruction in congenitally corrected transposition of the great arteries due to ventricular membranous septal aneurysms. *Circulation* 1976;54:679-83.
11. Marcelletti C, Maloney JD, Ritter DG, Danielson GK, McGoon DC, Wallace RB. Corrected transposition and ventricular septal defect: surgical experience. *Ann Surg* 1980;191:751-9.
12. Egloff L, Rothlin M, Schneider J, et al. Congenitally corrected transposition of the great arteries: a clinical and surgical study. *Thorac Cardiovasc Surg* 1980;28:228-32.
13. Williams WG, Suri R, Shindo G, Freedom RM, Morch JE, Trusler GA. Repair of major intracardiac anomalies associated with atrioventricular discordance. *Ann Thorac Surg* 1981;31:527-31.
14. Westerman GR, Lang P, Castaneda AR, Norwood WI. Corrected transposition and repair of associated intracardiac defects. *Circulation* 1982;66(Suppl):I197-202.
15. Hwang B, Bowman F, Malm J, Krongrad E. Surgical repair of congenitally corrected transposition of the great arteries: results and follow-up. *Am J Cardiol* 1982;50:781-5.
16. Metcalfe J, Somerville J. Surgical repair of lesions associated with corrected transposition: late results. *Br Heart J* 1983;50:476-82.
17. McGrath LB, Kirklin JW, Blackstone EH, Pacifico AD, Kirklin JK, Barger LM Jr. Death and other events after cardiac repair in discordant atrioventricular connection. *J THORAC CARDIOVASC SURG* 1985;90:711-28.
18. Di Donato RM, Wernovsky G, Jonas RA, Mayer JE Jr, Keane JF, Castaneda AR. Corrected transposition in situs inversus: biventricular repair of associated cardiac anomalies. *Circulation* 1991;84(Suppl):III193-9.
19. Huhta JC, Danielson GK, Ritter DG, Ilstrup DM. Survival in atrioventricular discordance. *Pediatr Cardiol* 1985;6:57-60.
20. Horvath P, Szufladowicz M, de Leval MR, Elliott MJ, Stark J. Tricuspid valve abnormalities in patients with atrioventricular discordance: surgical implications. *Ann Thorac Surg* 1994;57:941-5.
21. Graham TP Jr, Parrish MD, Boucek RJ Jr, et al. Assessment of ventricular size and function in congenitally corrected transposition of the great arteries. *Am J Cardiol* 1983;51:244-51.
22. Edwards WD. Embryology and pathologic features of Ebstein's anomaly. *Prog Pediatr Cardiol* 1993;2:5-15.
23. Lev M, Rowlett UF. The pathologic anatomy of mixed levocardia: a review of thirteen cases of atrial or ventricular inversion with or without corrected transposition. *Am J Cardiol* 1961;8:216-63.
24. Anderson RH, Arnold R, Wilkinson JL. The conducting system in congenitally corrected transposition. *Lancet* 1973;1:1286-8.
25. Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. *Circulation* 1974;50:911-23.
26. Bharati S, McCue CM, Tingelstad JB, Mantakas M, Shiel F, Lev M. Lack of connection between the atria and the peripheral conduction system in a case of corrected transposition with congenital atrioventricular block. *Am J Cardiol* 1978;42:147-53.
27. Lev M, Fielding RT, Zaeske D. Mixed levocardia with ventricular inversion (corrected transposition) with complete atrioventricular block: a histopathologic study of the conduction system. *Am J Cardiol* 1963;12:875-83.
28. Dick M II, Van Praagh R, Rudd M, Folkerth T, Castaneda AR. Electrophysiologic delineation of the specialized atrioventricular conduction system in two patients with corrected transposition of the great arteries in situs inversus {I,D,D}. *Circulation* 1977;55:896-900.
29. Wilkinson JL, Smith A, Lincoln C, Anderson RH.

- Conducting tissues in congenitally corrected transposition with situs inversus. *Br Heart J* 1978;40:41-8.
30. Huhta JC, Maloney JD, Ritter DG, Ilstrup DM, Feldt RH. Complete atrioventricular block in patients with atrioventricular discordance. *Circulation* 1983;67:1374-7.
 31. Russo P, Danielson GK, Driscoll DJ. Transaortic closure of ventricular septal defect in patients with corrected transposition with pulmonary stenosis or atresia. *Circulation* 1987;76(Suppl):III88-92.
 32. de Leval MR, Bastos P, Stark J, Taylor JFN, Macartney FJ, Anderson RH. Surgical technique to reduce the risks of heart block following closure of ventricular septal defect in atrioventricular discordance. *J THORAC CARDIOVASC SURG* 1979;78:515-26.
 33. Olinger GN, Maloney JV. Trans-pulmonary artery repair of ventricular septal defect associated with congenitally corrected transposition of the great arteries. *J THORAC CARDIOVASC SURG* 1977;73:353-7.
 34. Danielson GK. Surgical treatment of atrioventricular discordance. In: Losekoot TG, Anderson RH, Becker AE, Danielson GK, Soto B, eds. *Congenitally corrected transposition*. Edinburgh: Churchill Livingstone, 1983:177-90.
 35. Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC. Operative treatment of Ebstein's anomaly. *J THORAC CARDIOVASC SURG* 1992;104:1195-202.
 36. Benson LN, Burns R, Schwaiger M, et al. Radionuclide angiographic evaluation of ventricular function in isolated congenitally corrected transposition of the great arteries. *Am J Cardiol* 1986;58:319-24.
 37. Peterson RJ, Franch RH, Fajman WA, Jones RH. Comparison of cardiac function in surgically corrected and congenitally corrected transposition of the great arteries. *J THORAC CARDIOVASC SURG* 1988;96:227-36.
 38. Masden RR, Franch RH. Isolated congenitally corrected transposition of the great arteries. In: Hurst JW, ed. *Update III: the heart*. New York: McGraw-Hill, 1980:59-83.
 39. Parrish MD, Graham TP Jr, Bender HW, Jones JP, Patton J, Partain CL. Radionuclide angiographic evaluation of right and left ventricular function during exercise after repair of transposition of the great arteries: comparison with normal subjects and patients with congenitally corrected transposition. *Circulation* 1983;67:178-83.
 40. Dimas AP, Moodie DS, Sterba R, Gill CC. Long-term function of the morphologic right ventricle in adult patients with corrected transposition of the great arteries. *Am Heart J* 1989;118:526-30.
 41. Ilbawi MN, DeLeon SY, Backer CL, et al. An alternative approach to the surgical management of physiologically corrected transposition with ventricular septal defect and pulmonary stenosis or atresia. *J THORAC CARDIOVASC SURG* 1990;100:410-5.
 42. Di Donato RM, Troconis CJ, Marino B, et al. Com-

- bined Mustard and Rastelli operations: an alternative approach for repair of associated anomalies in congenitally corrected transposition in situs inversus {I,D,D}. *J THORAC CARDIOVASC SURG* 1992;104:1246-8.
43. Imai Y, Sawatari K, Hoshino S, Ishihara K, Nakazawa M, Momma K. Ventricular function after anatomic repair in patients with atrioventricular discordance. *J THORAC CARDIOVASC SURG* 1994;107:1272-83.
 44. Yagihara T, Kishimoto H, Isobe F, et al. Double switch operation in cardiac anomalies with atrioventricular and ventriculoarterial discordance. *J THORAC CARDIOVASC SURG* 1994;107:351-8.
 45. Brawn WJ, Wright JG, Sethia B, de Giovanni J, Silove ED. Atrial and arterial switch repair for corrected transposition and ventricular septal defect with severe tricuspid valve regurgitation [Abstract]. *Cardiol Young* 1993;3(Suppl):I58.

Discussion

Mr. Jaroslav F. Stark (*London, England*). I agree that in corrected transposition it is difficult or impossible to repair the valve. However, an alternative operative technique is now available: One can perform the so-called double switch: arterial switch and Mustard or Senning atrial repair, or, inasmuch as most of these patients have left ventricular outflow tract obstruction, a Rastelli operation plus Mustard or Senning. It is interesting how much the tricuspid incompetence can regress after this operation or how well it is tolerated once the tricuspid valve is placed into the pulmonary circulation.

Dr. van Son. Thank you very much for your kind remarks. I am well aware of the various repair techniques for corrected transposition that you just mentioned. The first variant, an atrial inversion procedure with Rastelli type of repair for corrected transposition, ventricular septal defect, and pulmonary stenosis or atresia, was first reported by Dr. Ilbawi and later modified by Dr. Imai in that he performed a direct anastomosis between the right ventricle and the distal pulmonary trunk (réparation à l'étage ventriculaire), thus avoiding the need for a conduit. Two Japanese groups, led by Dr. Imai and Dr. Kawashima, and Dr. Brawn from England subsequently reported performance of combined atrial inversion and arterial switch procedures for corrected transposition without pulmonary stenosis.

I think that the advantages of these operations, namely preservation of the morphologically left ventricle as the systemic ventricle and reduction or abolition of regurgitation of the morphologically tricuspid AV valve, must be balanced against the potential long-term adverse effects of the various components of these operations, namely development of supraventricular arrhythmias and pulmonary and systemic venous obstruction for the atrial inversion technique, potentially impaired pulmonary (morphologically left) ventricular function (Benson LN, Burns R, Schwaiger M, et al. *Am J Cardiol* 1986;58:319-24) for the arterial switch operation, and conduit obstruction for the Rastelli component.

Dr. Michel N. Ilbawi (*Oak Lawn, Ill.*). This work underlines two important principles that we have already

emphasized in the past. The first one is that the left-sided AV valve is abnormal and valve regurgitation should be treated aggressively and early to avoid ventricular dysfunction. This was not feasible in the past because the only surgical option was a valve replacement. However, recently we have used a variety of repair techniques for tricuspid valves and we have reported on five young patients with Ebstein-like anomalies, in whom these valvuloplasty techniques have worked very well so far, avoiding valve replacement in these children. I think it is important to emphasize that we should be aggressive with valve repair in tricuspid regurgitation at a very early age.

The second point, which Dr. Stark has already mentioned, is that the double switch operation is a very important operation in which the left ventricle becomes the systemic ventricle. Your data strongly support the concept that the right ventricle in the systemic circulation is bound to fail and therefore reinforces the concept behind the double switch operation.

Dr. Edward L. Bove (*Ann Arbor, Mich.*). May I take the liberty of asking you, Dr. Ilbawi, whether you think that is the operation of choice in any patient with corrected transposition and ventricular septal defect, even in the absence of AV valve regurgitation?

Dr. Ilbawi. Yes, I think so. We have enough data from our follow-up, as well as from the natural history of corrected transposition, to believe strongly that the right ventricle is bound to fail sooner or later. Therefore a double switch operation is strongly indicated as the primary operation of choice.

Dr. van Son. Although the concept of the combined atrial inversion and arterial switch operation is appealing, especially in patients with good pulmonary (morphologically left) ventricular function, the long-term results of this technique still need to be seen. I am mainly concerned about the atrial inversion component of this procedure. I emphasize that the key factor in the long-term outcome of patients with corrected transposition is *systemic ventricular function*; therefore, in the operation that we propose the regurgitant systemic AV valve needs to be replaced at an *early* stage to preserve systemic ventricular function.

Dr. Tom R. Karl (*Melbourne, Australia*). I would like to agree with Dr. Ilbawi and Mr. Stark. Our view has been

that even mild systemic AV valve insufficiency is usually a marker for right ventricular failure. If one wants to act early, one might consider a double switch operation for suitable patients. I would speculate that some of the problems related to atrial switch operations are caused or aggravated by failure of a systemic right ventricle. I believe that we will not see those problems in patients who have had a double switch, especially if a Senning operation is used for the atrial neoseptation.

In patients who are not suitable for a double switch, we would currently move toward a Fontan operation rather than an operation that septates the heart but leaves the right ventricle as the sole pump in the systemic circuit. With both approaches we have seen regression of moderate and, in some case, severe systemic AV valve insufficiency after the operation, without a specific procedure to repair or replace the valve. The results just presented do not justify early left AV valve replacement as an isolated procedure in this group of patients.

Dr. William G. Williams (*Toronto, Ontario, Canada*). Your data separated the ejection fractions above and below 44%. Beyond what limits would you no longer recommend a valve replacement and proceed to a heart transplantation?

Dr. van Son. During the last decade of our 30-year experience with systemic AV valve replacement in corrected transposition, we experienced a considerable improvement in early survival and encouraging late survival. This improvement is mainly due to increased awareness that timely replacement of the systemic AV valve is essential to preserve systemic (morphologically right) ventricular function. However, during the last decade we also operated on four patients who had severely impaired systemic ventricular function, all of whom were in NYHA functional class IV; three of these patients improved two functional classes, and one patient improved one class.

However, some patients with corrected transposition, systemic AV valve regurgitation, and severely impaired systemic ventricular function (it is difficult to provide a cutoff point in terms of ventricular ejection fraction) may be beyond corrective surgical relief, and for these patients heart transplantation may be the only viable option.