Intermediate Results From the Period of the Congenital Heart Surgeons Transposition Study: 1985 to 1989

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Background. The period of the Congenital Heart Surgeons Society (CHSS) study (1985 to 1989) provided a transition in the treatment of d-transposition of the great arteries. During this unusual time frame neonatal arterial switch (AS) and neonatal or late atrial baffle repair (Senning) were used in near equal proportion at one reporting institution. All the procedures were performed at this single institution, avoiding the variability intrinsic to a multicenter study. Intermediate follow-up of the results is presented.

Methods. During the period of the CHSS study, January 1985 to March 1989, 46 patients were enrolled in the CHSS study at one institution. Forty-four underwent either neonatal arterial (n = 14, 32%) or neonatal atrial (n = 19, 43%) or late atrial (n = 11, 25%) repair of d-transposition of the great arteries. Ages ranged from 4 to 80 days. Overall survival for the entire series was 91% (40/44). The survival of the AS group operated on in the neonatal period was 93% (13/14). The survival of the Senning group was 90% (27/30); late Sennings, 91% (10/11); and neonatal Sennings, 92% (12/13). Six neonatal Sennings were crossovers from the AS group with an 83% survival (5/6).

Results. Intermediate follow-up of 5.2 to 9.2 years revealed no late deaths. In the AS group there was no ventricular failure, no arrhythmias, and one reoperation for supravalvar pulmonic stenosis. In the Senning group, there was no ventricular failure, but significant complications developed in 10 patients: cardiac arrhythmias in 7, tachyarrhythmias requiring pharmacologic therapy in 4, and bradyarrhythmias in 3, 2 requiring permanent pacemaker insertion. Left ventricular outflow tract (subpulmonic) stenosis developed in 3 patients, 1 requiring a left ventricular to pulmonary artery conduit and permanent pacemaker. Systemic atrioventricular valve insufficiency has developed in 3 patients.

Conclusions. Results at one reporting institution from the CHSS study during this period of transition from late atrial repair (Senning) to neonatal atrial or arterial repair show comparable early mortality in all groups. However, the intermediate results at a mean of 6.7 years reveal fewer arrhythmic and functional complications in the AS group. The possibility of neonatal repair combined with low early and intermediate morbidity and mortality confirm AS as the treatment of choice for d-transposition of the great arteries.

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The period of the Congenital Heart Surgeons Society (CHSS) study, 1985 to 1989, provided a transition in the treatment of d-transposition of the great arteries (d-TGA) [1, 2]. During this unusual time frame both neonatal arterial switches and atrial baffle repairs were employed in a number of the study institutions. In several of these, late atrial baffle repairs were exclusively used, whereas in others the arterial switch was extensively employed. Meanwhile, several institutions were in a transition and in one, near-equal proportions of neonatal arterial switches, neonatal atrial baffle repairs (in many cases electively employed), and late atrial baffle repairs were reported as part of the study protocol. This represents a very unusual experience in neonatal transposition surgery, and the intermediate experience is explored [2].

Material and Methods

During the period of the CHSS study, January 1, 1985, to March 1, 1989, 46 patients were enrolled from a single reporting institution. All were patients less than 14 days of age at presentation to the reporting institution as defined by the CHSS study protocol. All other patients (ie, d-TGA with intact ventricular septum referred at more than 2 weeks of age for atrial repair, d-TGA with ventricular septal defect referred at more than 2 weeks of age for arterial reconstructions) were excluded from this evaluation. Treatment under the protocol was not randomly assigned or directed by the study, but rather "was selected by the physicians caring for the patient on the basis of their knowledge and experience." Forty-four of the 46 patients entered from our institution underwent either neonatal arterial (n = 14, 32%), neonatal atrial (n =

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19, 43%), or late atrial (n = 11, 25%) repair of d-TGA. There were 29 male and 11 female patients with ages ranging from 4 to 80 days. All were early and late procedures in the first 3 months of age. Three surgeons were involved in the treatment of the study group. Thirty-six procedures were performed by surgeon A, 4 by surgeon B, and 4 by surgeon C. All 14 arterial switches were performed by surgeon A. Two of the arterial switch patients had closure of associated ventricular septal defects. Follow-up is from 5.2 to 9.2 years (mean, 6.7 years). Follow-up data were available from the CHSS database on all 40 surviving patients, as well as current follow-up data from the individual cardiologists and direct family contact. Data on age at operation for each subgroup, height/weight plotted to age, and developmental issues were obtained for each patient, as well as mortality and morbidity. Intermediate follow-up included functional and anatomic defects, reoperations, pacemaker insertions, and rhythm disturbances, both within the entire cohort and subgroups, including neonatal arterial switches, neonatal elective atrial repairs and crossover atrial repairs, and late atrial repairs with results presented within the arterial switch and atrial repair, planned groups, and the three subgroups previously described. Statistical analysis involved χ^2 analysis, and Fisher exact test of small subgroups.

Results

During the period January 1985 to March 1989, 44 patients enrolled in the CHSS study with d-TGA were operated on by either an arterial switch or atrial repair with an overall survival of 91% (40 of 44 patients). No deaths occurred before surgical intervention.

Within the specific subgroups survival in the arterial switch group for the neonatal period was 93% (13 of 14), whereas survival of the Senning group was 90% (27 of 30), including 91% (10 of 11) of the patients in whom late Senning procedures were performed and (12 of 13) 92% of the patients undergoing elective neonatal Senning procedures [3]. Six patients having neonatal Senning procedures were crossovers from the arterial switch group with an 83% survival (5 of 6). A total of 88% (17 of 19) of the neonates having Senning procedures survived. No significant difference was noted between surgeons, and no significant difference was noted between the subgroups.

Table 1 demonstrates the results in relation to the planned groups of arterial switch and Senning procedures and the actual subgroups: arterial switch, crossover neonatal Senning, elective neonatal Senning, and late Senning. Overall mortality in the first (25 months) and second (25 months) halves of the study period is reported. All four deaths occurred among the 20 patients operated on in the first half of the study. These include the single arterial switch death (the only patient in whom arterial switch was performed in this period and the only one in whom arterial switch was performed without the single pulmonary arterial patch reconstruction technique who died of pulmonary arterial hemorrhage), as well as both neonatal Senning deaths (1 crossover, a single anterior coronary artery configuration and 1 elective, secondary to persistent pulmonary hypertension) from 10 such procedures performed during that period. The single late Senning death occurred among 9 such patients (a patient unique in this series due to presentation as an emergent respiratory infection with increasing cyanosis). No deaths occurred in the 24 patients operated on in the second half (25 months) of this series, including 13 arterial switch, 4 crossover neonatal Senning, 5 elective neonatal Senning, and 2 elective late Senning patients, demonstrating improved results not only in the arterial switch group but also the atrial subgroups, specifically the neonatal atrial repair in which little previous experience has been presented.

Table 1. Intermediate Results

Planned (n-death)	Actual (n-death)	1st Half (n-death)[a]	2nd Half (n-death)[a]
Arterial switch 20-2	Neonatal switch 14-1	1-1 (0)	13-0 (100)
	Neonatal Senning (crossover) 6-1	2-1 (50)	4-0 (100)
Atrial 24-2	Neonatal Senning 13-1	8-1 (87)	5-0 (100)
	Late Senning 11-1	9-1 (89)	2-0 (100)
Total 44-4		20-4 (80)	24-0 (100)

[a] Numbers in parentheses are percent survival.

Intermediate follow-up from 5.2 to 9.2 years (mean, 6.7 years) using the Kaplan-Meier actuarial method [4] reveals no late deaths in any group (Fig 1). Growth and development was normal in 32 including 22 of the 29 male patients and 10 of 11 female patients (Fig 2). Among the female patients in only 1 was growth less than the 25th percentile, and in this patient, less than the 5th percentile in an elective neonatal Senning patient. Among the male patients 6 were less than the 25th percentile, including 2 crossover Sennings and 1 elective neonatal Senning less than the 5th percentile, 1 elective neonatal Senning less than the 10th percentile, and 2 elective neonatal Senning patients less than the 25th

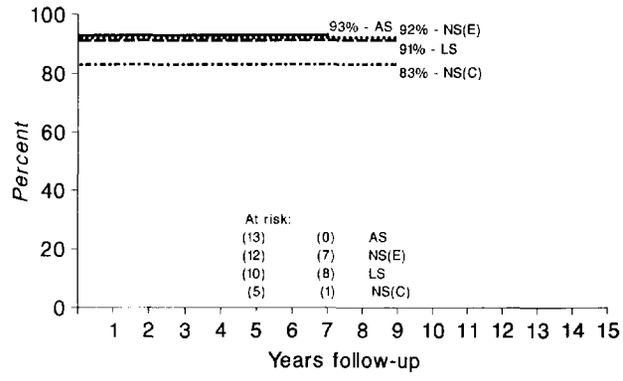

Fig 1. Kaplan-Meier actuarial survival. There is no significant difference between the subgroups. (AS = *arterial switch;* LS = *late Senning;* NS(C) = *crossover neonatal Senning;* NS(E) = *neonatal Senning [elective].*)

percentile. One late Senning and 1 arterial switch patient were at the 25th percentile for weight.

Neurologic problems were limited to one seizure disorder, which occurred after catheterization as an isolated occurrence 5 years postoperatively (a crossover Senning patient) and a learning disability in 1 patient (an elective late Senning patient). These findings are consistent with the age range of the patient group of less than 3 months (4 to 80 days), which has been demonstrated to decrease the risk of preoperative neurologic events, whereas improvement in perfusion techniques may have decreased the postoperative incidence of such problems [5].

Late morbidity can again be separated into the first and second halves of the study. In the arterial switch group there were no patients with ventricular failure, arrhythmias, or functional abnormalities. However, the greatest concern was that anatomic problems might develop. No significant supravalvar aortic stenosis and no coronary stenosis has been identified. One residual atrial septal defect has been followed up. There has been one reoperation for supravalvar pulmonic stenosis. This was repaired 2 years postoperatively with simultaneous repair of three small residual atrial septal defects. This patient in whom side-by-side great vessels were encountered at the original operation had a LeCompte maneuver performed and subsequent development of obstruction of the left pulmonary artery as it crossed anterior to an enlarged neoaorta (Fig 3). Two years postoperatively a patch of the left pulmonary artery was performed with successful resolution of the obstructive process. In 1 additional patient a mild supravalvar ridge is noted by echocardiography without a gradient and in 1 subvalvar mild pulmonic obstruction can be identified without a gradient.

Fig 3. Pulmonary arteriogram demonstrating left pulmonary artery stenosis secondary to compression by the dilated ascending aorta.

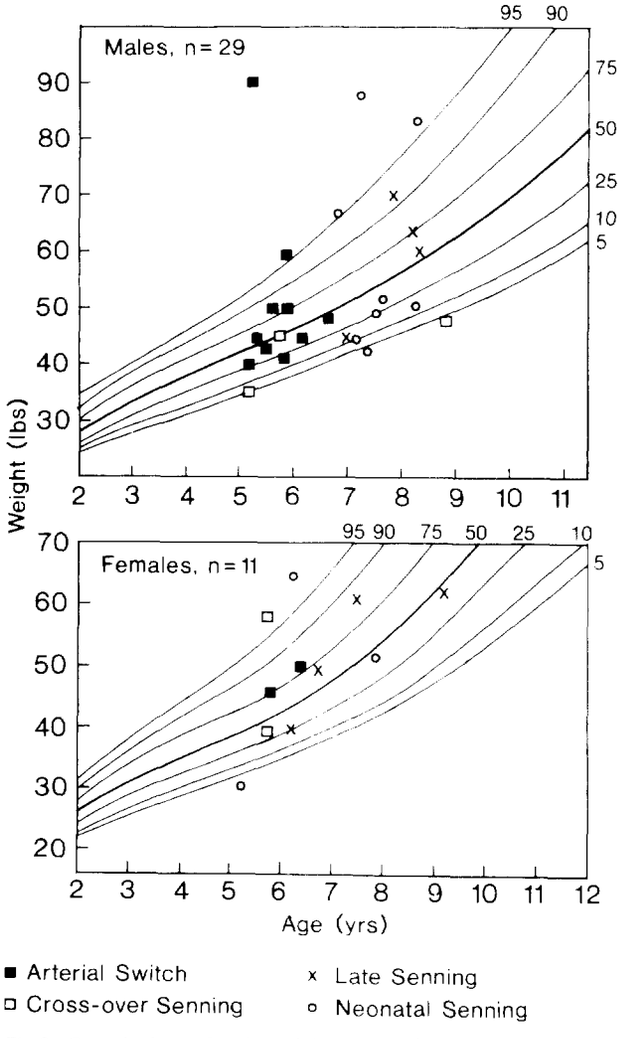

Fig 2. Growth chart of male and female weights.

In the Senning group no ventricular failure or anatomic problems such as baffle obstructions to the inferior vena cava or pulmonary veins have developed, even in the neonatal group. Only a single mild superior vena caval obstruction has been identified by echocardiography. However, significant complications have developed in 10 patients. Seven of these patients have cardiac arrhythmias. Tachyarrhythmias requiring pharmacologic therapy have developed in 4, and bradycardias have developed in 3, with 2 requiring permanent pacemaker insertion. Severe left ventricular outflow tract (subpulmonic) stenosis developed in 3 patients; 1 required a left ventricular to pulmonary conduit and permanent pacemaker, and in 2 abnormalities of the mitral valve, mitral stenosis, and mitral regurgitation are noted. Finally, most disturbingly, systemic atrioventricular valve (tricuspid valve) regurgitation has developed in 3 patients, although none of these has required operative intervention. Of note, although a higher mortality was observed among the first half of the patients in this series, in only 2 patients, both having neonatal Senning procedures (25%), have anatomic, functional, and arrhythmic complications been noted. In the latter period 12 complications were observed. Only one of these is of significance in the 13 neonatal arterial switches performed during

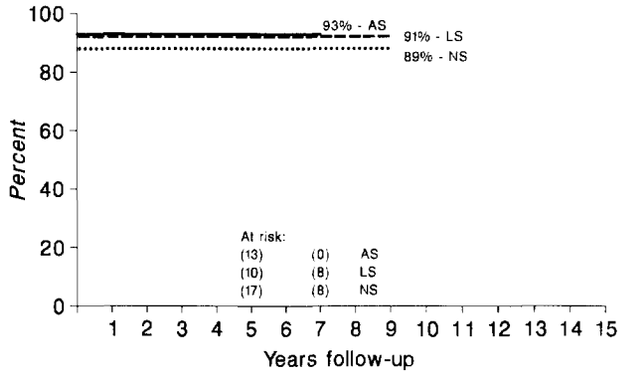

Fig 4. Kaplan-Meier actuarial survival combining the elective neonatal Senning and crossover neonatal Senning as no significant difference in intermediate follow-up was observed between these two groups. There is no significant difference between the subgroups. (AS = arterial switch; LS = late Senning; NS(E) = neonatal Senning [elective].)

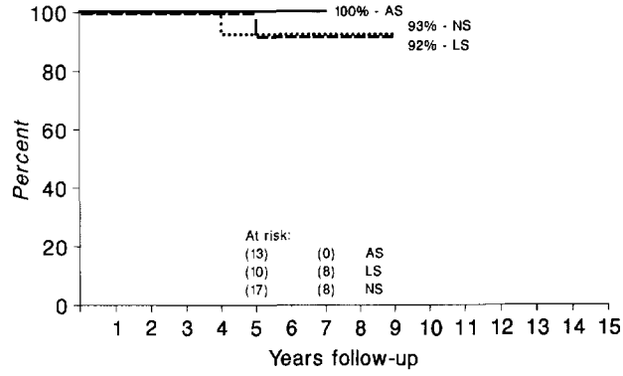

Fig 6. Kaplan-Meier actuarial freedom from pacemaker insertion. There is no significant difference between the subgroups. (AS = arterial switch; LS = late Senning; NS(E) = neonatal Senning [elective].)

that period, but they were present in 7 of the 11 Senning patients. The one left ventricular to pulmonary artery conduit was performed in a patient with a crossover Senning operation, and a permanent pacemaker insertion was necessary for complete heart block. A permanent pacemaker was placed in a neonatal Senning patient with bradyarrhythmia. The other 2 cases of severe subvalvar pulmonic stenosis and mitral stenosis are in patients with a neonatal crossover Senning and an elective neonatal Senning. Finally, systemic atrioventricular valve insufficiency is now beginning to be noted in both the late Senning and neonatal Senning population, a portent of future problems. Figure 4 demonstrates actuarial survival in the subgroups; arterial switch, late Senning, and neonatal Senning combining elective neonatal Senning and crossover neonatal Senning, because no significant difference was observed between the subgroups. Overall, Figure 5 demonstrates actuarial freedom from reoperation, Figure 6 actuarial freedom from pacemaker insertion, and Figure 7 demonstrates actuarial freedom from significant arrhythmia. Although again no significant difference was noted between the groups, this may be due to the small groups examined. Finally, new arrhythmic and functional problems are, however, surfacing in the Senning groups, most notably in the neonatal Sennings, while the neonatal arterial switch group remains to date without the potential anatomic complications, such as supravalvar aortic stenosis and coronary obstruction secondary to the basic technique or the arrhythmic and functional complications noted in the Senning group.

Comment

During the period of the CHSS study, January 1985 to March 1989, a transition point was experienced both nationally and in this reporting institution (a microcosm of this national process). Two forces were at work during this time frame, namely, the desire for neonatal repair of transposition and the specter of arterial reconstruction. Thus, multiple methods were used simultaneously and a comparison of their intermediate effectiveness is now possible.

The CHSS study produced a multiinstitutional database. The current study is drawn from the group of patients submitted by a single institution to that database

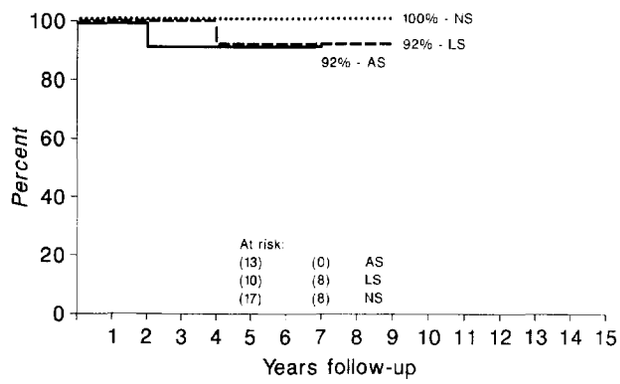

Fig 5. Kaplan-Meier actuarial freedom from reoperation. There is no significant difference between the subgroups. (AS = arterial switch; LS = late Senning; NS(E) = neonatal Senning [elective].)

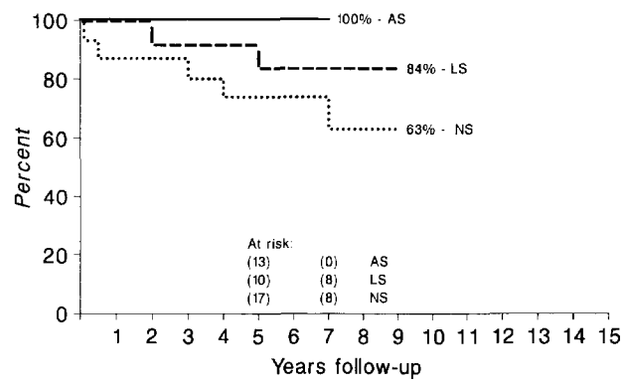

Fig 7. Kaplan-Meier actuarial freedom from arrhythmias. There is no significant difference between the subgroups. (AS = arterial switch; LS = late Senning; NS(E) = neonatal Senning [elective].)

under the protocol of the study. Multiinstitutional studies provide many positives, including the large group that can be accumulated [2]. However, such studies reflect the multiple centers from which the experience is drawn. In the absence of a planned protocol, multiple biases and levels of expertise can significantly affect the outcome data. Such a study is a global experience in a moment in time [1, 6–8]. The current study was performed at a single institution reporting to that study during that moment and represents a microcosm of the national process. It allows for detailed intermediate follow-up of near-equal groups of neonatal arterial switch, neonatal atrial baffle procedures (both from the elective and crossover groups), and late atrial baffle patients avoiding the variability intrinsic to the multicenter study, while providing follow-up of all three evolving methodologies.

The publications of the CHSS have focused on the evolving methodology of the arterial switch procedure including the early mortality, improvement in results experienced in many centers over the study period, technical problems of the technique, such as pulmonic stenosis and their effective solutions (as seen in the current study), and the lack of certain complications such as arrhythmias reflected in the current series. Likewise, the lack of certain anatomic complications such as supravalvar aortic stenosis and coronary stenosis are observed within the entire cohort. However, within the current study several observations concerning the atrial subgroups are evident. Neonatal atrial repairs represented a small sideshow in the total experience of the CHSS study and as such were not examined in detail. In the study's publications, atrial repair mortality was noted to be stable without improvement over time. Although this may be true of the elective late atrial repairs, it was at variance with the current findings in the neonatal atrial patients. Elective neonatal atrial repairs were uncommon in the larger study group, but represented a significant percentage in the current series and did improve over time. Likewise, crossovers were noted to carry an increased mortality in the initial CHSS study of 50% (2 of the 4 patients), and in the subsequent review in 1992 of 513 cases, 14 were crossovers with 4 deaths. Of note, in that initial series, 2 of the patients were from the current study group with the single death (a patient with microcephaly and single anterior coronary artery) representing the only crossover death in this series. Subsequently, no crossover deaths occurred among the 4 crossover neonatal atrial repairs performed in the second half of this study. Of the total of 14 crossovers reported in the study of 1992, 6 were from this study group with 5 survivors, whereas only 5 of the 8 crossovers from other institutions survived. Thus, like arterial switch patients of the current series, atrial repairs in the neonatal group demonstrated improvement in survival over the study period: whereas the mean initial survival of the multistudy group was 82%, ours was 91% at the conclusion of the study period, reflecting improvement over time both in the arterial switch subgroup and the neonatal atrial repairs. Intermediate follow-up of both the atrial and arterial switch subgroups demonstrate no continued late risk of mortality (hazard function) to date, although functional and rhythm complications have become evident in the atrial but not the arterial repair groups [8–11].

Thus, the results of the collaborative efforts of the CHSS show a number of consistencies and several inconsistencies with this microcosm of that study. First, in the case of late Senning operations a large prior experience allowed for a standard systematic approach to atrial repair of d-TGA at more than 1 month of age, which is reflected in the results of the general study [9]. However, elective neonatal Senning operations represent an unusual approach with a learning curve and improvement in operative mortality over time. Likewise, crossover neonatal Senning operation through the experience generated in the elective group demonstrated a reduced mortality over time. Both approaches, however, have demonstrated increasing morbidity, and this is concerning in the intermediate follow-up. Although mortality can be reduced, the neonatal Senning procedure has been associated with increased risk in this experience of arrhythmias, functional and anatomic problems, and a suggestion of impaired growth and development. In contrast, the arterial switch procedure performed within the protocol of the CHSS study in the current study group demonstrated a 93% late survival. There was a single incidence of significant and, we believe now, preventable anatomic abnormality requiring reoperation, and there have been no significant functional or arrhythmic complications. No supravalvar aortic stenosis, no coronary arterial stenosis, no subpulmonic stenosis, and no atrioventricular valve regurgitation has occurred.

The current study, in which all three modalities were used simultaneously, demonstrated that the arterial switch procedure resulted in comparable early and late survival and to date has "outperformed" the other method employed in avoiding anatomic, functional, and arrhythmic complications. These intermediate results confirm the neonatal arterial switch as the procedure of choice in d-TGA.

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DISCUSSION

DR BRUNO MESSMER (Aachen, Germany): I am somewhat amazed that you did not find any rhythm disturbances in your cohort of arterial switch patients. We have studied the late postoperative rhythm behavior in 33 of 78 patients who had neonatal switch repair. Supraventricular and sporadic ventricular extrasystoles were quite common on Holter electrocardiograms. More dangerous rhythm disturbances such as persistent junctional rhythm or supraventricular tachycardia, however, were present in only 3 patients, which is roughly 10%. Rhythm problems are certainly less after an arterial switch than after the Senning or Mustard procedure, but they are not absent as one would assume after your presentation. Therefore I would like to ask you whether you checked your patients with long-term electrocardiograms or only by looking at casually recorded electrocardiograms.

DR TURLEY: This material was from the Congenital Heart Surgeons Society database. It did not include Holter data unless the individual cardiologists had chosen to perform that study. We have followed up 74 patients with arterial switches in our entire series and have noted 3 patients in whom rhythm disturbances have developed in that total group. The series presented today is a very unusual group of patients within an unusual time frame followed up by the database, and in that very small group, no rhythm disturbances were seen.

DR LUDWIG VON SEGESSER (Zürich, Switzerland): Did you evaluate the patients studied with regard to aortic incompetence, and is there a difference between groups?

DR TURLEY: We have reviewed all of our arterial switch patients by echocardiography and have seen only mild incompetence in 3 of the patients in the current series.

DR CARL L. BACKER (Chicago, IL): I enjoyed your presentation. We reviewed our patients at Children's Memorial Hospital and did a similar comparison [1]. We looked at neonates with transposition of the great arteries and assessed 23 infants who underwent a Mustard operation and 37 an arterial switch procedure. Our findings were almost identical to yours; there was a significantly higher incidence of both early and late arrhythmias after the Mustard operation (atrial repair). The other factor that we reviewed and that I was curious as to whether you had any data on is related to the higher incidence of tricuspid regurgitation after the Senning procedure. We looked at ventricular function angiographically, comparing both systemic ventricular ejection fraction and systemic ventricular end-diastolic volume in both the arterial switch (left ventricle) and the Mustard groups (right ventricle). The right ventricular function was significantly lower after the Mustard operation as compared with the left ventricular function after the arterial switch. The systemic ventricular end-diastolic volume was significantly greater after the Mustard procedure as compared with the arterial switch. Would you comment on the significance of tricuspid regurgitation after the Senning procedure, and ventricular function, if that was examined?

DR TURLEY: Concerning the issues of tricuspid regurgitation and ventricular function after the atrial repairs, we have been disturbed by the findings in 3 patients of regurgitation noted in the past 2 years. Before that time, no regurgitation had developed in the series. Several years ago, we reported our follow-up on Mustard operations performed on infants less than 3 months of age. This patient series now extends to 18 years of follow-up, and when we have compared them with arterial switch patients in exactly the functional categories we are discussing, we found that right ventricular performance was significantly impaired, even though those patients have continued to experience excellent clinical courses. There were no early deaths in that group of 36 Mustard patients less than 3 months of age and only one late death, actually a drowning; however, major rhythm disturbances and the need for pacemakers were noted and those rhythm disturbance episodes increased over time. Now only 48% of those patients are without major rhythm disturbances, and we believe the findings in the current study using the Senning procedure for atrial repair in neonates and infants demonstrate these same findings in relation to both the need for pacemaker and rhythm disturbances. We project the same problems as seen in our much longer Mustard experience.

DR JOHN E. MAYER, JR (Boston, MA): Could I ask just one question. Are these all intact ventricular septums?

DR TURLEY: During the period of the Congenital Heart Surgeons Society study we enrolled 46 patients; in 2 with ventricular septal defects, Rastelli repair was performed. Two of the patients with arterial switches presented today, likewise, had simultaneous closure of the ventricular septal defect at the time of neonatal arterial switch. These are included as no difference in their outcomes from the remaining arterial switch patients was evident. No ventricular septal defect repairs were performed in the atrial groups.

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