JACC Vol. 6, No. 1
July 1985:221–7

PEDIATRIC CARDIOLOGY

Prevention of Sudden Death After Repair of Tetralogy of Fallot: Treatment of Ventricular Arrhythmias

ARTHUR GARSON, JR., MD, FACC, DAVID C. RANDALL, PAUL C. GILLETTE, MD, FACC, RICHARD T. SMITH, MD, JEFFREY P. MOAK, MD, PAT McVEY, RN, DAN G. McNAMARA, MD, FACC

Houston, Texas

The majority of sudden deaths after repair of tetralogy of Fallot have been presumed to be due to ventricular arrhythmia; however, it remains to be demonstrated that antiarrhythmic medication reduces the incidence of sudden death. Since 1978, ventricular arrhythmias have been treated aggressively; these include any ventricular arrhythmia on routine electrocardiogram and more than 10 uniform premature ventricular complexes per hour on 24 hour electrocardiogram. A review was undertaken of 488 patients followed up for more than 1 month after repair of tetralogy of Fallot (mean follow-up time 6.1 years); 13.5% had ventricular arrhythmia on routine electrocardiogram. Ventricular arrhythmia appeared from 2 months to 21 years postoperatively (mean 7.3 years). Ventricular arrhythmias were significantly ($p < 0.01$) related to: longer follow-up duration, older age at follow-up, older age at operation and higher postoperative right ventricular systolic and end-diastolic pressures. Ventricular arrhythmia on routine electrocardiogram occurred in 100% of those who later died suddenly compared with 12% of those who did not die ($p < 0.01$).

Treatment for ventricular arrhythmia was given to 46 patients and considered "successful" if there were fewer than 10 uniform premature ventricular complexes per hour on 24 hour electrocardiogram. A successful drug was found in 44 of the 46: 30 of 34 given phenytoin, 6 of 9 given propranolol, 1 of 7 given quinidine, 1 of 2 given disopyramide, 8 of 9 given mexiletine and 4 of 5 given amiodarone. Sudden death did not occur in any of the 44 patients with successful antiarrhythmic treatment compared with a 39% incidence of sudden deaths in those with ventricular arrhythmias who were untreated (7 of 21) or had unsuccessful antiarrhythmic treatment (2 of 2) ($p < 0.01$). In conclusion, with aggressive antiarrhythmic treatment of ventricular arrhythmias, sudden death in postoperative tetralogy of Fallot has been significantly reduced.

(J Am Coll Cardiol 1985;6:221–7)

Sudden death occurs more commonly after repair of tetralogy of Fallot than in any other condition currently encountered by pediatric cardiologists (1). This problem has been identified for more than 10 years (2–5) and its cause has been the subject of much speculation. Since our observation in 1979 (6) that more than 30% of those with premature ventricular complexes on routine electrocardiogram after repair of tetralogy of Fallot died suddenly, we have aggressively treated ventricular arrhythmias in these patients. In this report, we review the outcomes of all of our patients who have undergone repair of tetralogy of Fallot with specific reference to the effect of treatment of ventricular arrhythmias.

Methods

Patients. The medical records of all patients examined in the Section of Pediatric Cardiology, Texas Children's Hospital who had intracardiac repair of tetralogy of Fallot were reviewed. Patients were included only if a follow-up time of more than 1 month was available. All "routine" electrocardiograms (12 lead with rhythm strip) and 24 hour electrocardiograms taken more than 1 month after the patient's operation were reviewed for the presence and type of ventricular arrhythmias, as well as any conduction disturbance. The time of first appearance of these abnormalities was recorded. The most severe ventricular arrhythmia on

From the Lillie Frank Abercrombie Section of Cardiology, Department of Pediatrics, Baylor College of Medicine and Texas Children's Hospital, Houston, Texas. Dr. Garson is an Established Investigator of the American Heart Association, Dallas, Texas. This study was supported in part by Grant HL07190 from the National Institutes of Health, United States Public Health Service, Washington, D.C., Grant RR00188 from the Department of Health and Human Services, United States Public Health Service and a grant from the J.S. Abercrombie Foundation, Houston, Texas. Manuscript received November 13, 1984; revised manuscript received January 21, 1985, accepted February 21, 1985.

Address for reprints: Arthur Garson, Jr., MD, Pediatric Cardiology, Texas Children's Hospital, 6621 Fannin, Houston, Texas 77030.

0735-1097/85/$3.30

JACC Vol. 6, No. 1
July 1985:221–7

the routine electrocardiogram or 24 hour electrocardiogram was also noted.

Postoperative catheterization. Routine postoperative hemodynamic cardiac catheterization was recommended to all patients. It was performed in the postabsorptive, sedated state with meperidine (2 mg/kg body weight) and promethazine (1 mg/kg) administered intramuscularly as premedication, with intravenous use of reduced dosages as necessary throughout the procedure. Right ventricular peak systolic and end-diastolic pressures and aortic peak systolic pressure were recorded as was the presence of a residual ventricular septal defect with a greater than 1.5:1 ratio of pulmonary to systemic blood flow. Because pulmonary artery angiography was not routinely performed, pulmonary insufficiency was not evaluated. The majority of our patients received a transanular patch during the repair of tetralogy of Fallot and so pulmonary insufficiency was a necessary consequence. Because all right ventricular angiograms were not available and the written estimate of ventricular contractility was subjective, right ventricular systolic function was not assessed in this study. In any patient who underwent two cardiac catheterizations, the most recent data were recorded, unless the patient underwent a second operation. In this case, the postoperative data from the first operation were used.

Follow-up. In addition, the age and duration from initial repair to the most recent follow-up were recorded. In terms of follow-up status, sudden death was defined as an event that occurred outside the hospital in a patient who was not acutely ill. This sudden event rendered the patient unconscious and resulted in death without regain of consciousness (7).

Treatment of ventricular arrhythmias. Beginning in 1978, all patients after repair of tetralogy of Fallot were treated with antiarrhythmic medication if there were any premature ventricular complexes on routine electrocardiogram or more than 10 uniform premature ventricular complexes in any hour (or any multiform premature ventricular complexes, couplets or ventricular tachycardia) on 24 hour electrocardiogram. Before 1978, a systematic approach to treatment was not used.

Only one antiarrhythmic drug was used at a time and it was not used in combination with any other cardiac drug except digoxin or diuretic drugs, or both. For each drug, the dosage was increased until side effects occurred or the serum concentration was well above the accepted therapeutic range. In general, treatment began with phenytoin (8) (after a loading dose, maintenance 5 to 7 mg/kg daily), and if this was not effective or side effects developed, recently mexiletine was given (9) (9 to 15 mg/kg daily). Next, if the right ventricular contractility was thought to be normal, propranolol (2 to 4 mg/kg daily) was administered. In a few patients, quindine (30 to 40 mg/kg daily) was administered (10). Finally, if all conventional agents failed, amiodarone (11) (after a loading dose, 5 mg/kg daily) was given.

Drug effectiveness was assessed by 24 hour electrocardiogram, obtained at least every 6 months. A drug was considered effective if there were no episodes of ventricular tachycardia, couplets or multiform premature ventricular complexes and there were fewer than 10 uniform premature ventricular complexes per hour on two consecutive 24 hour electrocardiograms, as well as all follow-up 24 hour electrocardiograms. Treatment has been continued indefinitely because the ventricular arrhythmias have returned in those who have voluntarily stopped their medication.

Statistical analysis. Comparison of grouped data was done by chi-square. Comparison of continuous data from two groups was done by Student's t test and comparison of data from three or more groups was done by analysis of variance with specific comparisons of two subgroups made after Bonferroni's modification (12). Statistical significance was inferred if the probability of the event occurring by chance was less than 0.05.

Results

Clinical Features

Age at repair. The study group consisted of 488 patients, each of whom had been followed up for more than 1 month after repair of tetralogy of Fallot. The average age at repair was 6.4 years (range 3 months to 36 years). In 5% of our patients, repair was performed before 1 year of age, in 52% before 5 years of age and in 1.7% before 20 years of age.

Hemodynamics. Cardiac catheterization was performed in 342 (70%) of the 488 patients. The postoperative right ventricular systolic pressure ranged from 17 to 120 mm Hg (mean 48.5). In 25% of these patients, the right ventricular systolic pressure was 60 mm Hg or more. The right ventricular end-diastolic pressure ranged from 1 to 30 mm Hg (mean 7.6); in 44% of the patients, it was 8 mm Hg or more. A residual ventricular septal defect with a pulmonary to systemic flow ratio greater than 1.5:1 was present in 21 (6.1%).

Ventricular arrhythmias. Premature ventricular complexes were noted on at least one routine electrocardiogram in 67 (13.7%) of the 488 patients; in 58 (11.9%) these were uniform and in 9 (1.8%) they were multiform. These arrhythmias were first noted on the electrocardiogram between 2 months and 21 years after repair (mean 7.3 years). In only 18% of those with premature ventricular complexes was the arrhythmia noted within the first postoperative year. Premature ventricular complexes first appeared more than 10 years postoperatively in more than one-third of the patients with this arrhythmia. Since each of the 488 patients underwent a routine electrocardiogram on each clinic visit, this incidence of ventricular arrhythmias is not biased by selection.

However, a 24 hour electrocardiogram was obtained in only 97 (20%) of the patients, and the selection of patients

for such a test might have been biased by the presence of ventricular arrhythmia on routine electrocardiogram, palpitation or syncope. It is only recently that all patients have undergone a 24 hour electrocardiogram as part of a regular postoperative visit. Of the 97 patients who underwent a 24 hour electrocardiogram, ventricular arrhythmia was found in 71 (73%). In 56% of these patients, there were uniform premature ventricular complexes, in 8% multiform complexes, in 3% couplets and in 6% ventricular tachycardia.

Drug treatment. Antiarrhythmic drugs (other than digitalis) were administered to 46 (10%) of the patients. An "effective" drug according to the previously listed criteria was found in 44. This control was achieved with an average of 1.4 drugs per patient (range 1 to 5). The average duration of treatment was 2.3 years (range 6 months to 6.2 years).

Phenytoin was used most frequently and was successful in 88% of the patients (Table 1). In 12% of the patients, a generalized pruritic rash appeared within 10 days of beginning treatment; therefore, the drug was discontinued. One patient died suddenly while supposedly on phenytoin treatment. He had not undergone a 24 hour electrocardiogram in more than 2 years and his compliance had been questionable. Quinidine treatment was only successful in one the the seven patients to whom it was given, and disopyramide in one of the two patients. Propranolol treatment was successful in six of the nine patients, including one successfully managed through a normal pregnancy, labor and delivery.

Of the investigational drugs, we gave mexiletine to nine patients after repair of tetralogy of Fallot, and the drug was successful in eight. In the remaining patient, the drug had to be discontinued because it caused a rash. The average duration of treatment with mexiletine was 1.4 years (maximum 2.5). We gave amiodarone to five patients and had success in four. The average duration was 2.6 years (maximum 3.5). Although the duration of therapy remains short, there have been no major side effects caused by amiodarone. Corneal microdeposits were found in two patients, and one had nausea that responded to a decrease in dosage.

Conduction disturbance. The combination of complete right bundle branch block and left anterior hemiblock was found in 52 patients (10.6%). There was no statistical association between the presence of premature ventriuclar complexes and right bundle branch block with left anterior hemiblock. There were no instances of late-appearing second or third degree atrioventricular (AV) block on routine electrocardiogram. In 2 of the 97 patients who underwent a 24 hour electrocardiogram, short episodes of type I second degree AV block were noted when the patients were asleep.

A pacemaker was implanted in eight patients. In four, the pacemaker was placed in the immediate postoperative period for surgically acquired complete AV block at the time of the repair. In two patients, second degree AV block persisted for long periods of time after the operation. A pacemaker was implanted in one of these two (6 months postoperatively) because of the electrophysiologic finding of Mobitz type II second degree AV block with block below the bundle of His on intracardiac recordings, and in the other (6 years postoperatively) because of reduced exercise tolerance associated with a slow ventricular rate. In the two remaining patients, a pacemaker was implanted for sinus bradycardia, one had reduced exercise tolerance and one had coexistent atrial flutter and developed profound bradycardia after quinidine was administered for control of his tachyarrhythmia.

Follow-up. On follow-up study, 475 patients (97%) were alive. Sudden death had occurred in nine (1.8%) and four patients died nonsuddenly from cardiac causes (congestive heart failure in two, bacterial endocarditis in one and one during reoperation). The sudden deaths occurred at an average age of 16.5 years (range 5 to 25). This was from 6 months to 16 years postoperatively (mean 5.9 years). The remaining 475 patients were last seen at an average age of 12.4 years (range 9 months to 38 years). This was an average of 6.1 years postoperatively (range 2 months to 23 years). Follow-up data within the last 5 years were available for 248 (52%) of the patients.

Factors predisposing to ventricular arrhythmias. *Hemodynamics.* Because of the possible bias introduced by selection of patients for 24 hour electrocardiography, only the routine electrocardiogram was used for these comparisons. The results of cardiac catheterization were compared between the groups of patients with and without any premature ventricular complexes on routine electrocardiogram. Those with such complexes had a significantly higher right ventricular systolic pressure (57.8 ± 3.2 [mean ± SE] versus 47.0 ± 1.1, $p < 0.001$). In general, the higher the right ventricular systolic pressure, the greater the number of patients with premature ventricular complexes, the greatest prevalence occurring with a right ventricular systolic pressure greater than 60 mm Hg (Fig. 1). The right ventricular end-diastolic pressure was also related, but less significantly, to premature ventricular complexes. Such complexes were found in 10.6% of those with a right ventricular end-diastolic pressure of 7 mm Hg or less compared with 18.4% of those with an end-diastolic pressure of 8 mm Hg or more ($p < 0.05$). Since the right ventricular systolic and

Table 1. Treatment of Ventricular Arrhythmias in 46 Postoperative Patients

Drug	No. of Trials*	% Success	Side Effects (%)
Phenytoin	34	88	Rash (12)
Propranolol	9	67	Lethargy (11)
Quinidine	7	14	0
Disopyramide	2	50	0
Mexiletine	9	89	Nausea (22); rash (11)
Amiodarone	5	80	0

*Since patients may have received more than one drug, the total number of trials is greater than the total number of patients.

JACC Vol. 6, No. 1
July 1985:221-7

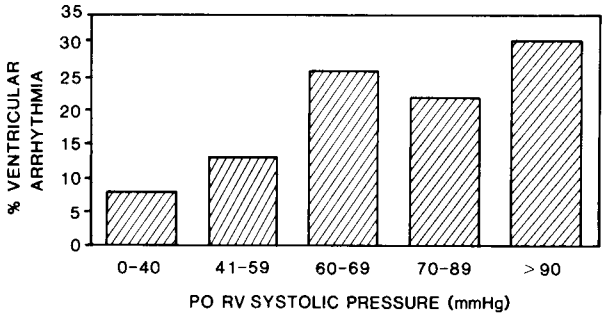


Figure 1. Ventricular arrhythmias related to postoperative (PO) right ventricular (RV) systolic pressure.

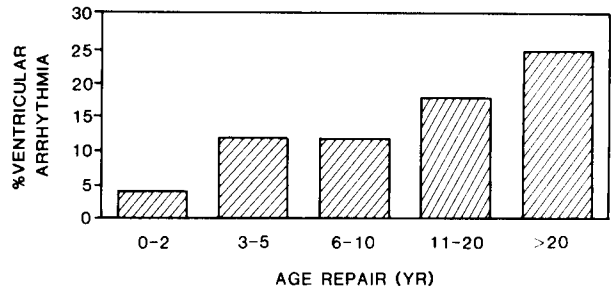


Figure 3. Ventricular arrhythmias related to age at repair.

end-diastolic pressures are strongly related, the comparison was made of premature ventricular complexes in patients with a right ventricular systolic pressure less than 60 mm Hg. In this subgroup, if the right ventricular end-diastolic pressure was 8 mm Hg or more, 14% had premature ventricular complexes compared with 8% of those with a lower end-diastolic pressure ($p = 0.09$). Therefore, there is a trend toward an independent effect on frequency of premature ventricular complexes if the right ventricular end-diastolic pressure is high when the right ventricular systolic pressure is low. Using the same analysis, when the right ventricular systolic pressure is high, the right ventricular end-diastolic pressure does not have an independent additive effect on premature ventricular complexes.

Timing of surgery and follow-up. The presence of premature ventricular complexes was strongly related to the duration of follow-up. Patients with such complexes had a significantly longer follow-up (7.6 ± 0.7 years) than those without (5.8 ± 0.2 years) ($p < 0.01$). With increasing age at follow-up, there was a steadily increasing incidence of premature ventricular complexes; among patients 30 years of age and older, one-third had such complexes on routine electrocardiogram (Fig. 2).

Age at the time of surgical repair was related to the presence of premature ventricular complexes (Fig. 3). With increasing patient age at repair, there was an increased incidence of later development of premature ventricular complexes. Further analysis, however, indicates that the duration of follow-up was probably the more significant factor, since age at repair and duration of follow-up are significantly related. The major reason for this is that in recent years (with shorter follow-up time), the age of repair has decreased. For patients operated on before 2 years of age, the mean follow-up time is only 3.1 years. Since the average duration postoperatively of the development of premature ventricular complexes is 7 years, on the average these patients with early repair have not lived long enough to develop premature ventricular complexes. In addition, among those few patients operated on before 2 years of age who have been followed up for 10 or more years, 50% have premature ventricular complexes.

Factors predisposing to sudden death. *Right ventricular pressure.* All the hemodynamic factors (right ventricular systolic pressure, end-diastolic pressure and ventricular septal defect) that predispose to premature ventricular complexes were also significantly higher in patients who later died suddenly than in those who did not. Of the nine patients who died suddenly, eight had undergone postoperative cardiac catheterization. In six, the right ventricular systolic pressure was 60 mm Hg or more *and* the end-diastolic pressure was 8 mm Hg or more. In two patients, only the end-

Figure 2. Ventricular arrhythmias related to age at follow-up.

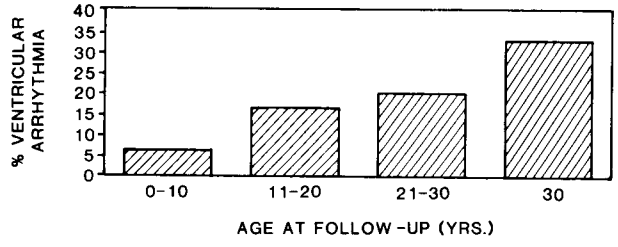
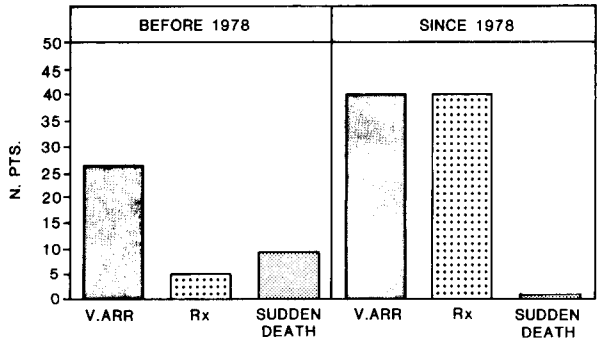


Figure 4. Comparison of aggressive antiarrhythmic treatment since 1978 with previous years. The number of patients with ventricular arrhythmia (V.ARR) on routine electrocardiogram are compared with those treated (Rx) and those who died suddenly. Before 1978, few patients were treated and a significant number died. Since 1978, the incidence of ventricular arrhythmias has increased but all patients have been treated. Only one patient died suddenly and he was thought to be noncompliant.



diastolic pressure increased. The age at repair was also significantly related to sudden death. Among those who died suddenly, only one (11%) of nine had repair before 5 years of age compared with those who did not die suddenly, among whom 53% had repair before 5 years of age. Additionally, seven of nine patients who died suddenly had the operation before 1974, when the routine use of cold cardioplegia during cardiopulmonary bypass was instituted.

Ventricular arrhythmia. A major factor in sudden death was the presence of premature ventricular complexes on routine electrocardiogram. Each of the nine patients who died suddenly had premature ventricular complexes on routine electrocardiogram taken during a clinic visit before death. Among patients who did not die suddenly, premature ventricular complexes occurred in 12% ($p < 0.0001$).

Role of antiarrhythmic treatment. In assessing the prevention of sudden death, we compared the incidence of sudden death in patients who were and were not treated for premature ventricular complexes. Among patients who were not treated, 33% died suddenly compared with 4% of those who were treated with antiarrhythmic drugs ($p < 0.01$). There were no significant differences between those who received treatment and those who did not with respect to age at repair (7.5 ± 1.0 versus 9.4 ± 1.0 years, $p = 0.52$). However, if the stringent criteria are considered for documented control of ventricular arrhythmia on 24 hour electrocardiogram, we have had no deaths among the 44 patients treated properly compared with 9 sudden deaths (39% mortality rate) in the 23 patients who either were not treated or who did not have successful control ($p < 0.001$) (Table 2). From 1960 through 1977 before the aggressive treatment program began, there were eight sudden deaths and since 1978 there has been only one in a patient who was being treated but who was probably noncompliant (Fig. 4).

To assure that the treatment of patients since 1978 was a major factor in the reduction in sudden death, it is important to compare those who died suddenly with an appropriate control group. We compared the 9 patients who died suddenly with the 44 patients with premature ventricular complexes who are alive and receiving treatment. There was no difference in any variable including hemodynamic status, age at surgery or age at follow-up (Table 3). Therefore, the only difference between those who died and those who are alive is that those who are alive were treated.

Table 2. Sudden Death and Ventricular Arrhythmia in 488 Postoperative Patients

	Sudden Death (no.)	Alive (no.)
No. of ventricular arrhythmias	0	421
Ventricular arrhythmias		
No treatment	7	14
Treatment unsuccessful	2	0
Treatment successful	0	44
Total	9	479

Discussion

Prevention of sudden death with antiarrhythmic drugs. Using an aggressive treatment program aimed at abolition of premature ventricular complexes from the routine and 24 hour electrocardiograms, we have demonstrated that sudden death after repair of tetralogy of Fallot can be markedly reduced. It has taken many years to demonstrate this effect because, fortunately, sudden deaths have been rare and usually occurred late after surgery. Therefore, it required a similar duration of follow-up in treated patients to be sure that it was the antiarrhythmic therapy that was causing the decrease in such deaths.

The incidence of sudden death in untreated patients after repair of tetralogy of Fallot will now be difficult to determine. All the early reports (13,14) noted an incidence of sudden death of approximately 5%. However, because more recent reports have shown an increasing incidence of ventricular arrhythmias with increasing duration of follow-up, it is likely that without treatment of these patients now, the incidence of late sudden death would be considerably higher than 5% in another 5 years of follow-up study. Therefore, it is possible that in 1985, had patients been left untreated, those who were 15 to 20 years postoperative might have an incidence of sudden death considerably higher than 5%.

It is possible that the implantation of a pacemaker in some of our patients may also have contributed to the reduction in late death. However, the number of these patients is small compared with that of patients with ventricular arrhythmias. It is difficult to determine the contribution of more modern surgical techniques, such as the use of cardioplegia, to prevention of sudden death. In our series, as well as others (14), patients who had cardiopulmonary bypass with cardioplegia have later died suddenly.

Drug treatment protocol. Compared with adult studies (15,16) on ventricular arrhythmias and sudden deaths, the ventricular arrhythmias in our patients were rather easily controlled. Our patients required only 1.4 drugs per patient for control compared with 4 to 5 drugs per patient in many adult studies (15,16). In addition, the spectrum of drugs that are effective in our patients is quite different from that in adults. Phenytoin and mexiletine in adults are not recognized as the best primary antiarrhythmic drugs, whereas in our patients they were effective in 85 to 90%. Propranolol, also not a traditional primary agent for ventricular arrhythmias in adults, was relatively effective in the postoperative patients with tetralogy to whom it was given (that is, only those with good ventricular function). Quinidine, traditionally a widely used ventricular antiarrhythmic agent in adults, was relatively unsuccessful in our patients. Fi-

Table 3. Fifty-three Patients With Premature Ventricular Complexes: Correlation With Other Factors

	Sudden Death (no. = 9)*	Treated and Alive (no. = 44)*	p Value
Postop RVSP (mm Hg)	66.5 ± 7.7	60.3 ± 4.6	0.50 (NS)
Postop RVEDP (mm Hg)	8.9 ± 1.0	7.3 ± 0.8	0.32 (NS)
Age at repair (yr)	10.2 ± 1.1	7.5 ± 1.0	0.16 (NS)
Follow-up duration (yr)	5.2 ± 1.7	8.0 ± 4.8	0.17 (NS)
Age at last follow-up (yr)	15.5 ± 2.0	14.7 ± 1.3	0.76 (NS)

*For those undergoing cardiac catheterization, no. = 8 for sudden death group and no. = 28 for treated group. NS = no statistical difference; p = probability; postop = postoperative; RVEDP = right ventricular end-diastolic pressure; RVSP = right ventricular systolic pressure.

nally, amiodarone therapy seems to be as successful in children as it is in adults. The reason for the differences in ease of control and type of drugs that achieve control are not known at present. It is possible that in adults with coronary artery disease, intermittent ischemia may alter the blood supply to certain arrhythmogenic areas causing electrophysiologic changes in the tissue and reducing the amount of antiarrhythmic drug delivered to that tissue. It is also possible that the cellular mechanisms for ventricular arrhythmias are different in patients after repair of tetralogy of Fallot than in those with coronary artery disease. This is under active investigation in our experimental laboratory (17).

Criteria for drug treatment. Fortunately, the long-term side effects of drug treatment in our patients have been minimal, and actually less than those found in adults. It is clear that, using our recommendations for treatment of even frequent uniform premature ventricular complexes on 24 hour electrocardiogram, we are overtreating a certain group of our patients. It would, indeed, be desirable to develop stricter criteria for treatment of ventricular arrhythmias. We (18) and others (19,20) have performed ventricular extrastimulus protocols on such patients in an attempt to identify those who require treatment. At present, the clinical implications of inducibility of ventricular arrhythmias are uncertain, since even sustained ventricular tachycardia has been induced in asymptomatic patients with no ventricular arrhythmia on Holter monitor (19).

Ideally, a randomized placebo-controlled prospective study should be undertaken to determine which arrhythmias need treatment. This is, in a sense, being done, since at present there are numerous treatment strategies around the world. Through the auspices of the Pediatric Electrophysiology Society, we plan to review our results periodically. With the psychological and economic burdens imposed by long-term drug therapy, it would be desirable to develop a more direct approach to the prevention of sudden death.

Primary prevention of sudden death. Since it appears that the majority of sudden deaths are related to ventricular arrhythmias, the question arises as to whether ventricular arrhythmias can be prevented primarily. There is a statistical relation between right ventricular systolic pressure and ventricular arrhythmias, but this is clearly not the entire answer since 10% of patients with a low pressure have ventricular arrhythmias and 70% of those with a high pressure do not. Nonetheless, it might be that if the hemodynamic results could be improved, the incidence of ventricular arrhythmias might decrease. In certain cases it is not possible to reduce the right ventricular outflow obstruction any further because of small distal pulmonary arteries. However, there are increasing numbers of reports (13,14) linking ventricular arrhythmias to abnormal right ventricular function (which we could not examine in this retrospective review). This is likely to be related to pulmonary insufficiency. From our study and others, we know that the incidence of ventricular arrhythmias increases with the increase in duration of follow-up. We know that right ventricular dysfunction may develop over time and it may be this dysfunction with its attendant stretch in areas adjacent to the ventriculotomy that may provide the basis for the development of late ventricular arrhythmias. In the future, primary prevention of ventricular arrhythmias will likely be directed toward 1) production of as small an area of ventriculotomy and outflow resection as possible to give a small scar, 2) reduction of the right ventricular systolic pressure, and 3) insertion of some device (monocusp, valve or otherwise) that will prevent pulmonary regurgitation. Although a step has been made in the prevention of sudden death, if we could prevent the conditions for development of ventricular arrhythmias these children could be permitted to lead an active life without the need for drugs.

Conclusions. The majority of sudden deaths after repair of tetralogy of Fallot are due to ventricular arrhythmias that occur in the setting of abnormal hemodynamics and increasing time after surgery. With aggressive use of antiarrhythmic drugs and adequate control of ventricular arrhythmia, the incidence of sudden death can be significantly reduced.

References

1. Garson A, McNamara DG. Sudden death in a pediatric cardiology population, 1958–1983: relation to prior arrhythmias. J Am Coll Cardiol 1985;5:134B–7B.
2. Gillette PC, Yeoman MA, Mullins CE, McNamara DG. Sudden death after repair of tetralogy of Fallot: electrocardiographic and electrophysiologic abnormalities. Circulation 1977;56:566–70.
3. Quattlebaum TG, Varghese PJ, Beill CA, Donahoo JS. Sudden death among postoperative patients with tetralogy of Fallot. Circulation 1976;54:289–96.
4. James FW, Kaplan S, Chou TC. Unexpected cardiac arrest in patients after surgical correction of tetralogy of Fallot. Circulation 1976;54:671–9.
5. Wolff GS, Rowland TW, Ellison RC. Surgically induced right bundle branch block with left anterior hemiblock. Circulation 1972;45:587–93.
6. Garson A, Nihill MR, McNamara DG, Cooley DA. Status of the adult and adolescent after repair of tetralogy of Fallot. Circulation 1979;59:1232–40.
7. Lambert EC, Menor VA, Wagner HR, Vlad P. Sudden unexpected death from cardiovascular disease in children. Am J Cardiol 1974;34:89–96.
8. Garson A, Kugler JD, Gillette PC, Simonelli A, McNamara DG. Control of late postoperative ventricular arrhythmias with phenytoin in young patients. Am J Cardiol 1980;46:290–4.
9. Moak JP, Gillette PC, Garson A. Mexiletine. An alternative to dilantin for pediatric ventricular arrhythmias (abstr). Circulation 1984;70 (suppl II):II-207.
10. Garson A. Evaluation and treatment of chronic ventricular dysrhythmias in the young. Cardiovasc Rev 1981;2:1164–82.
11. Garson A, Gillette PC, McVey P, et al. Amiodarone treatment of critical arrhythmias in children and young adults. J Am Coll Cardiol 1984;4:749–55.
12. Wallenstein S, Zucker C, Fleiss JL. Some statistical methods useful in circulation research. Circ Res 1980;47:1–12.
13. Deanfield JE, McKenna WJ, Hallidie-Smith KA. Detection of late arrhythmia and conduction disturbance after correction of tetralogy of Fallot. Br Heart J 1980;44:248–53.
14. Kavey REW, Blackman MS, Sondheimer HM. Ventricular dysrhythmias after repair of tetralogy of Fallot: incidence and severity. Am Heart J 1982;103:342–50.
15. Spielman SR, Schwartz JS, McCarthy DM, et al. Predictors of success or failure of medical therapy in patients with chronic recurrent sustained ventricular tachycardia: a discriminant analysis. J Am Coll Cardiol 1983;1:401–11.
16. Mason JW, Winkle RA. Electrode catheter arrhythmia induction in the selection and assessment of antiarrhythmic drug therapy for recurrent ventricular tachycardia. Circulation 1978;58:971–80.
17. Garson A. Ventricular dysrhythmias after congenital heart surgery: a canine model. Pediatr Res 1984;18:1112–20.
18. Garson A, Porter CJ, Gillette PC, McNamara DG. Induction of ventricular tachycardia during electrophysiology study after repair of tetralogy of Fallot. J Am Coll Cardiol 1983;1:1493–502.
19. Vetter VL, Horowitz LN. Electrophysiologic results of repair of tetralogy of Fallot (abstr). Am J Cardiol 1982:49:999.
20. Kugler JD, Mooring PK, Pinsky WW, Cheatham JP, Hofschire PJ. Sustained ventricular tachycardia following repair of tetralogy of Fallot: new electrophysiologic findings (abstr). Am J Cardiol 1982;49:998.