

# Status of Congenital Heart Disease Patients 10 to 15 Years After Surgery

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**T**HE MEDICAL rehabilitation program of the New York State Department of Health provides services to State residents under 21 years of age who are crippled by accidents, congenital malformations, birth injuries, or debilitating diseases to help them increase their capacity for education or remunerative employment. The program is administered jointly by the local and State health departments, and each county is reimbursed by the State for 50 percent of the costs.

Although this program has been underway for 40 years, evaluations of the surgical and rehabilitative procedures for specific conditions have been made to determine only short-term physical restoration and vocational and educational attainment. However, the investment of considerable expenditures in hospitalization, surgery, prosthetics, and physical and other therapies justifies a closer examination of the results and the long-term progress of the patients.

Evaluation of these procedures from a medical standpoint is often difficult because there are no accepted criteria for measuring clinical improvement. In most conditions, however, it is possible to make clinical judgments as to whether or not surgical procedures have helped the patient. The medical elements of the rehabilitation process should not be limited to the physical and surgical rehabilitative procedures, but should include followup and determination of whether the patients are in their proper school grade or employed. Unfortunately,

long-term followup procedures are seldom included in the rehabilitation programs of health departments or other agencies providing similar services to children and adults.

In Erie County, the department of health conducted a long-term followup of 100 patients with cerebral palsy and an evaluation of a group of children with cleft palate (1, 2). In 1954, Wallace and associates studied the first 129 children with congenital heart disease admitted to the New York City medical rehabilitation program (3). They examined the cost and the kinds of services given these children, with emphasis on the implications of the findings for improving services. Data related to the physical status of these patients, however, were limited to a short time after surgery. Other studies have emphasized the postoperative clinical status of patients without an evaluation of their educational and employment status.

Surgery for certain types of congenital heart disease has been performed in hospitals in Buffalo, N.Y., since 1947. But, in the late forties, a limited number of congenital anomalies were amenable to cardiac surgery. As surgery was limited to closed heart procedures in 1947-52, a common congenital cardiac defect, the inter-ventricular septal defect, was not included in this program at that time (4, 5). Referrals were made to the Buffalo hospitals from Erie County and other western New York counties. Most of the patients were referred to the Children's Hospital of Buffalo, the major cardiac center.

The study reported here was undertaken in 1961-62 to evaluate the effectiveness of surgical correction of congenital cardiac anomalies, using such measurements as mortality, postoperative clinical status, educational attainment, and pres-

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ent occupation, for 94 patients who had undergone surgery 10-15 years previously. Of these patients, 85 had been admitted to the Children's Hospital of Buffalo and 9 to the Buffalo General Hospital from 1947 through 1952. The study also included an analysis of the medical rehabilitation costs, by type of heart defect and nature of the services performed.

### Methods

Members of the cardiology departments at both hospitals reviewed the hospital records as well as the records of the cardiac diagnostic clinics which the patients attended. Many of the Erie County patients were seen and followed in a special cardiac diagnostic clinic administered and financed by the county's health department and staffed by cardiologists of the Children's Hospital of Buffalo.

At the time of this study, 68 patients survived. Of these, 30 were seen at the special cardiac diagnostic clinics, and none required further surgery. Another 15 patients were seen at clinics held specifically to determine their present clinical status; 3 required further surgery and all improved after surgery. Of 23 patients not seen during the study year, 8 were patients with patent ductus arteriosus who could not be located and who, by definition, were considered completely rehabilitated. Ten patients who did not keep their clinic appointments or who could not because they lived out-of-state had either patent ductus arteriosus or coarctation, and one patient had a corrected aortic septal defect. All of these 11 patients were also, by definition, completely rehabilitated. The remaining four patients were three with tetralogy of Fallot and one with a coarctation who could not be located. These four patients were classified according to their clinical status at the time of their last visit.

Educational and employment information was obtained for 56 patients by one of the following methods:

1. Home visit by a caseworker of the health department (36 patients, 64 percent).

2. Personal visit or telephone conversation by a public health intern with residents of counties other than Erie County within a 30-mile radius of the city of Buffalo (8 patients, 14 percent).

3. Use of mail to contact persons in the remainder of New York State or living in other States (12 patients, 22 percent).

The cost of services for each patient was obtained from the business office of the Erie County Health Department or the bureau of medical rehabilitation of the New York State Department of Health. Death certificates were reviewed to obtain adequate information as to the cause of death.

Definitions were established as to the medical rehabilitation status of the patients with special reference to the four major cardiac defects. These standards facilitated a practical evaluation of each patient's current status and enabled us to classify each case in one of the following three groups.

### *Complete Rehabilitation*

*Tetralogy of Fallot.* Presence of a continuous murmur. Asymptomatic and able to attend school or work full time.

*Patent ductus arteriosus.* Disappearance of a continuous murmur for not less than 5 years. Able to attend school or work full time. Electrocardiogram and chest X-ray show no abnormality.

*Coarctation.* Return of blood pressure to normal within 5 years. Able to attend school or work full time. Electrocardiogram and chest X-ray show no abnormality. (No data on vascular changes in optic fundi. This information would be helpful.)

*Pulmonary stenosis.* Electrocardiogram shows no, or only slight, right ventricular hypertrophy or a reduction of right ventricular pressure to less than 50 mm. Hg. Able to attend school or work full time. Chest X-ray shows no abnormality; moderately loud to loud murmur still present.

*Other cardiac defects.* Aortic septal defect completely rehabilitated on disappearance of murmur. Able to attend school or work full time. Electrocardiogram and chest X-ray show no abnormality.

### *Partial Rehabilitation*

*Tetralogy of Fallot.* Continuous murmur with some symptomatic improvement. Able to attend school or work part time.

**Table 1. Distribution of patients, by medical rehabilitation status and type of heart defect**

Medical rehabilitation status	Tetralogy of Fallot		Patent ductus arteriosus		Coarctation		Pulmonary stenosis		Other defects		Total	
	Number	Percent	Number	Percent	Number	Percent	Number	Percent	Number	Percent	Number	Percent
Complete.....	14	45	32	97	7	70	5	72	1	8	59	63
Partial.....	4	13	0	---	1	10	1	14	0	---	6	7
No improvement.....	2	6	0	---	0	---	0	---	1	8	3	3
Died.....	11	36	1	3	2	20	1	14	11	84	26	27
Total.....	31	---	33	---	10	---	7	---	13	---	94	---

<sup>1</sup> One patient with a pulmonary valvulotomy in 1952, but no data on rehabilitation status following open heart repair in California, 1954.

*Coarctation.* Some reduction in blood pressure. Able to attend school or work part time.

*Pulmonary stenosis.* Physically improved, but less than "complete" rehabilitation. Able to attend school or work part time.

*No Improvement*

Clinical status remains essentially the same as preoperative status.

**Findings**

At the time of their first surgical experience, the 94 patients (56 females and 38 males) ranged in age from 3 months to 20 years. All but 22 resided in Erie County. By 1962, 26 (27 percent) of the 94 patients had died as a result of surgery or of a complication of their cardiac anomaly.

In terms of current clinical status, after one or more surgical procedures 59 (63 percent) of the patients were considered completely rehabilitated, 6 (7 percent) partially rehabilitated, and 3 (3 percent) showed no improvement (table 1). The entire group of 94 patients had experienced 126 operative procedures, exclusive of catheterization (table 2). The average case was followed 8.8 years.

Because of the differences among heart anomalies, the medical rehabilitation results which follow are based on the anatomical cardiac defect.

*Tetralogy of Fallot.* Of 31 patients in this group, ranging in age at first operation from 10 months to 20 years, 18 (58 percent) were rehabilitated. Seven (58 percent) of the 12 operated on at the age of 3 years or younger died.

Four (21 percent) of the 19 patients first operated on after the age of 3 years died postoperatively. At present, further operations have been performed on 4 of the 5 living patients first operated on before the age of 3 years, whereas only 5 of the 15 living patients first operated on after the age of 3 years have required further surgery.

*Patent ductus arteriosus.* Of the 33 patients in this group, 32 were completely rehabilitated. The one death in the group was that of a patient with a recurrent ductus.

*Coarctation.* Eight of the 10 patients in this group, ranging in age at first operation from 3 months to 20 years, were rehabilitated. A 3-month-old infant and a 19-year-old female died.

*Pulmonary stenosis.* Ages at first operation of the patients in this group ranged from 4 to 11

**Table 2. Distribution of patients, by type of heart defect, number of years followed, and number of operative procedures**

Type of defect	Number of patients	Average number of years followed postoperatively <sup>1</sup>	Number of operative procedures <sup>2</sup>
Tetralogy of Fallot.....	31	9.3	50
Patent ductus arteriosus.....	33	8.3	36
Coarctation.....	10	8.8	11
Pulmonary stenosis.....	7	9.2	15
Other defects.....	13	10.5	14
Total.....	94	8.8	126

<sup>1</sup> Excluding deaths immediately following surgery.

<sup>2</sup> Excluding cardiac catheterizations.

years. The only death in the group was that of a 6-year-old boy. Of the other six patients, five were completely and one partially rehabilitated.

*Other cardiac defects.* This group included 13 patients with the following malformations: 2 with complete transposition of the great vessels, 2 with tricuspid atresia with a rudimentary right ventricle, 2 with dextrocardia with pulmonary atresia, 2 with aortic septal defect, 3 with ventricular septal defect, 1 with subvalvular aortic stenosis, and 1 with truncus arteriosus. Seven of these patients underwent cardiac surgery at less than 1 year of age in a desperate attempt to save their lives. Of the two surviving patients in this group, one is completely rehabilitated following surgery for an aortic septal defect. The second is a boy operated on for a misdiagnosed patent ductus arteriosus whose postoperative diagnosis was a ventricular septal defect. He has not undergone surgery for this defect as he has been doing well. This patient has been classified as "no improvement" and is being followed in the cardiac clinic.

As to the educational and employment status of 56 of the 68 surviving patients, 53 (95 percent) are presently in their appropriate school grade or are gainfully employed as workers or housewives (table 3). The other 5 percent includes three mentally retarded patients. Educational and employment data on 12 patients not included in this table were not available because the patients had moved from their last known address and had no forwarding address.

The educational and employment status information was related to age distribution in the

five cardiac defect groups. This revealed no significant differences between the groups except for a higher percentage, 63 percent, of employment in the coarctation group, compared with 33 percent in the pulmonary stenosis group. This difference is probably due to a present mean age in the coarctation group of 25 years, and 88 percent of this group was first operated on after the age of 9 years. The present mean age of the pulmonary stenosis patients is 18 years.

In the group of 17 patients classified as "employed" in table 3, 7 are housewives and 1 is a hospital volunteer worker. Briefly, the remaining nine patients include two secretaries earning between \$2,000 and \$4,000 a year, and a third secretary earning between \$4,000 and \$6,000 a year. The fourth is a skilled worker earning more than \$6,000 a year and the father of two children. The fifth, a father of one child, earns between \$4,000 and \$6,000 a year and is pursuing higher education in night school. The sixth is a mechanic who earns between \$4,000 and \$6,000 a year and has one child. Two others earn between \$2,000 and \$4,000 a year: one is an apprentice tool and die maker, and the other is an accounting clerk. No information was available as to the income of the ninth patient, a 30-year-old single man, who is working away from home. Ten of these 17 patients have a total of 20 children.

The total expenditures were \$78,117 for all services received by the 94 patients in this study, from the time of their first operation (1947-52) to the present time (tables 4 and 5). The total average cost per patient was \$822, while the

**Table 3. Educational and employment status of 68 surviving patients, by type of heart defect**

Type of heart defect	School status		Employed	Unknown	Total
	Full-time student <sup>1</sup>	Mentally retarded			
Tetralogy of Fallot.....	11	1	5	3	20
Patent ductus arteriosus.....	18	0	6	8	32
Coarctation.....	2	0	5	1	8
Pulmonary stenosis.....	3	2	1	0	6
Other defects.....	2	0	0	0	2
Total.....	<sup>2</sup> 36	3	<sup>3</sup> 17	12	68

<sup>1</sup> All but two of these children are in their appropriate school grade.

<sup>2</sup> Includes three students with part-time jobs earning less than \$2,000 per year.

<sup>3</sup> Includes seven housewives.

**Table 4. Distribution of expenditures, by type of heart defect and medical services received**

Type of heart defect	Hospital	Physician services	Special nursing	Catheterization	Other costs	Total
Tetralogy of Fallot.....	\$13,302	\$10,893	\$2,676	\$401	\$1,075	\$28,347
Patent ductus arteriosus.....	10,304	8,479	2,200	553	1,085	22,621
Coarctation.....	3,821	2,349	543	0	430	7,143
Pulmonary stenosis.....	5,845	3,393	918	581	217	10,955
Other.....	3,784	3,668	766	428	405	9,051
Total.....	37,056	28,782	7,103	1,963	3,212	78,117

average cost per patient to the county was \$686, exclusive of insurance benefits and contributions from parents. Hence, 84 percent of the expenditures for 94 patients was assumed by the medical rehabilitation program. This cost per patient figure of \$822 closely approximates the average cost of \$900 per patient for closed heart surgery, as reported by Norval (6).

**Discussion**

*Medical.* At present, based on the postoperative medical, educational, and employment status of the 94 patients comprising this study, 70 percent are rehabilitated. This is both a rewarding and impressive finding, as the patients were followed an average of 8.8 years postoperatively, and rehabilitation was based on the physical status of the patients in conjunction with their ability to attend school or work full time or part time.

Between 80 and 97 percent of the 50 patients in the patent ductus arteriosus, coarctation, and pulmonary stenosis groups were rehabilitated

as a result of surgery (table 1). This percentage of rehabilitation is probably due to the successful techniques of surgical correction for these defects.

The lower figure of 58 percent rehabilitation in the tetralogy of Fallot group may be attributed to the fact that the corrective surgical procedures for this anomaly were palliative rather than curative. Seven of the 11 deaths were of patients under the age of 3 years. This concentration of mortality in the younger age group appears to be due to the severity of illness in these children who required emergency surgery.

In the "other" cardiac defect group, the importance of age is again evident as a contributing factor to the death of 11 of the 13 patients. As previously indicated, 7 (54 percent) of these patients were operated on before the age of 1 year in a desperate attempt to save their lives. Fortunately, since the advent of open heart surgery there has been a decreasing mortality in younger children undergoing surgical correction of congenital cardiac defects (7).

*Social.* The educational and employment information on 56 of the 68 surviving patients indicates the substantial benefits to be gained by the surgical rehabilitation of children with congenital heart disease. Medical rehabilitation was obtained in 53 (95 percent) of these patients when measured by their physical status and educational or employment attainments.

*Financial.* The higher cost per patient in the tetralogy of Fallot and the pulmonary stenosis groups may be explained by the fact that the majority of these patients had more than one operation. In our experience, patients with patent ductus arteriosus had the lowest per patient cost since they required only one opera-

**Table 5. Average cost per patient, by type of heart defect and postoperative result**

Type of heart defect	Average cost per patient		
	Total	Surviving	Deceased
Tetralogy of Fallot.....	\$914	\$1,048	\$675
Patent ductus arteriosus.....	665	669	535
Coarctation.....	714	658	939
Pulmonary stenosis.....	1,369	1,735	272
Other.....	754	1,164	672
Total average cost per patient.....	822	885	666

tion. Their hospital stay was short, and only a few required cardiac catheterization in order to establish the diagnosis.

In all but the coarctation group, the average cost per living patient exceeded the average cost per deceased patient. This is attributed to the fact that many of the deceased patients had relatively short periods of hospitalization compared with the surviving patients; approximately one-third of the surviving patients required further operative procedures and hospitalization. The reverse was true in the coarctation group because the two deceased patients had poor postoperative courses. They died in the hospital 1 and 2 months after surgery.

### Summary

Of 94 patients who underwent surgery for the first time between 1947-52 for correction of congenital heart anomalies, 65 (70 percent) were considered medically rehabilitated 10-15 years later, 3 showed no improvement, and 26 died. All the patients were followed postoperatively an average of 8.8 years.

Information on 56 of the 68 surviving patients revealed that 95 percent were either attending school on a grade level commensurate with their ages or were housewives or gainfully employed workers.

All the patients received treatment in Erie County under the New York State medical rehabilitation program. The average expendi-

ture per patient, based on both living and deceased patients, was \$822, of which 84 percent was paid by the New York State Health Department and the county in which the patient lived. The remaining 16 percent was financed by insurance benefits or contributions from parents, or both.

### REFERENCES

- (1) Mosher, W. E.: An evaluation of young adults with severe neuromuscular disorders to determine community needs for vocational rehabilitation services. *Amer J Public Health* 49: 622-633 (1959).
- (2) Mosher, W. E.: Review of 96 cleft palate children, aged 5-10, who had received rehabilitative services through the medical rehabilitation program. Erie County Health Department, New York, 1955. Mimeographed.
- (3) Wallace, H. M.: Congenital heart disease in a medical rehabilitation program. *J Pediat* 45: 273-284 (1954).
- (4) Chenoweth, A. D., and Saffion, S.: Children with congenital heart disease served in regional centers, 1952-1956. *Amer J Public Health* 75: 377-386, May 1960.
- (5) Piero, M., et al.: Congenital malformations of the heart and great vessels. *JAMA* 183: 109-112, Jan. 26, 1963.
- (6) Norval, M. A.: Minnesota's regional cardiac program. *Public Health Rep* 77: 311-314, April 1962.
- (7) Cooley, D. A., and Ochsner, J. L.: Surgical treatment of cardiovascular anomalies in infants less than one year of age. *Heart Bull* 11: 12-17, January-February 1962.

## Gehrig Heads Bureau of Medical Services

Dr. Leo J. Gehrig, deputy chief of the Bureau of Medical Services in the Public Health Service since July 1962, assumed the duties of chief on April 1, 1964. He succeeded Dr. James V. Lowry, who has retired from the Service and is now director of the California Department of Mental Hygiene.

Dr. Gehrig, a career officer in the Service since 1945, was the first medical director of the U.S. Peace Corps. A native of Duluth, Minn., he received his B.S. and M.D. degrees from the University of Minnesota. After special training in tuberculosis he became acting director of tuberculosis control in Alaska. Later he served in the Public Health Service Hospitals in Seattle, Wash., and Staten Island, N.Y., as chief of chest surgery and deputy chief of general surgery. In 1957 he became deputy chief of the PHS Division of Hospitals.