CONGENITAL HEART DISEASE

Fifth USA-USSR Joint Symposium

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FIFTH USA-USSR JOINT SYMPOSIUM

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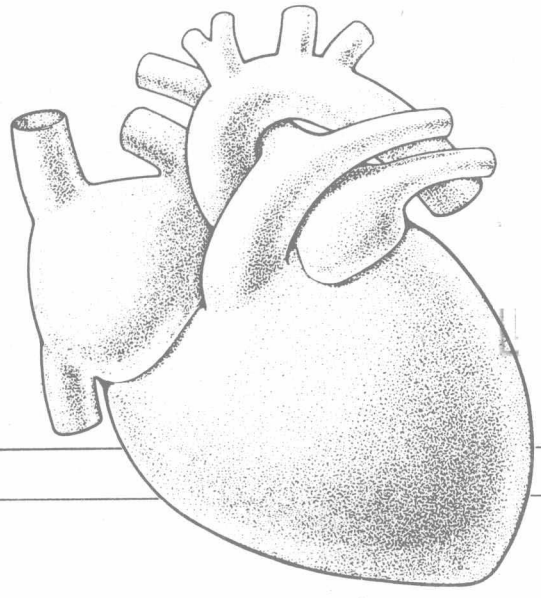

PREFACE

The Fifth US-USSR Joint Symposium on Congenital Heart Disease was held May 9-10, 1983, in Williamsburg, Virginia. These bilateral symposia are held in accordance with an agreement between the governments of the United States and the Soviet Union in the field of medical science and public health. The meetings have alternated between the USA and the USSR, giving the participants the opportunity to visit and learn about the research facilities and medical clinics in each country. The proceedings of the symposia have been published in English and in Russian.

Congenital heart disease is a serious disease among infants and children in both countries. The purpose of US-USSR collaboration in this area is to exchange information on new methods of diagnosis and postoperative care in order to further reduce mortality from this disease and to improve the surgical treatment of complex heart defects. At the fifth symposium the joint activities of more than 10 years were discussed. Since this bilateral effort was initiated in 1972, 10 delegations of scientists have met at joint symposia and working meetings to exchange research findings and new approaches, 24 fellows have worked in each other's laboratories for extended periods of time, and two collaborative research projects have been completed and published.

Collaborative activities in this area have resulted in an expanded network of information exchange for the diagnosis, treatment, and followup care of patients with congenital heart disease. The 18 papers presented in these proceedings are indicative of the range and depth of problems faced in this research area. It is hoped that the joint discussions and exchange of ideas will contribute to solutions to these disease problems.

The US coordinator for this problem area is Dr. Henry T. Bahnson, Professor and Chairman, Department of Surgery, University of Pittsburgh School of Medicine, Pittsburgh, Pennsylvania; the USSR coordinator is Academician Vladimir I. Burakovsky, Professor of Surgery and Director of the A.N. Bakulev Institute for Cardiovascular Surgery, USSR Academy of Medical Sciences, Moscow, USSR.

On behalf of the National Heart, Lung, and Blood Institute, I wish to thank Dr. Bahnson and Academician Burakovsky, Mrs. Zena McCallum, who serves as the Institute liaison, the US and USSR participants, and all those in both the USSR and USA who participated in the planning of this symposium and the editing of these proceedings.

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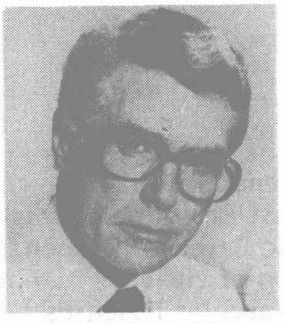

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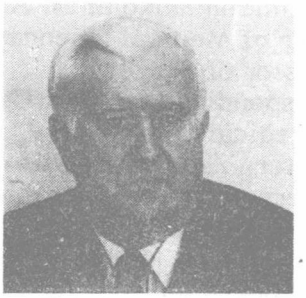

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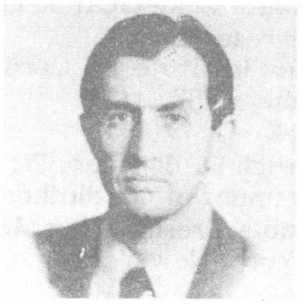

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ANOMALOUS ORIGIN OF THE CORONARY ARTERY FROM THE PULMONARY ARTERY - ANALYSIS OF TREATMENT EXPERIENCE AND LATE RESULTS

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ABSTRACT

This paper concerns a rare heart disease--anomalous origin of the coronary arteries from the pulmonary trunk. Twenty-two patients ranging in age from 2 months to 39 years were treated and followed up. They included 20 patients with anomalous origin of the left coronary artery and 2 with anomalous origin of the right coronary artery from the pulmonary artery. Sixteen patients underwent 17 operations. On one patient with anomalous right coronary artery, the artery's ostium was reimplanted in the aorta, and plastic surgery was performed to correct an interventricular septal defect. A second patient underwent patch closure of an aortopulmonary septal defect with interposition of the right coronary ostium. The pericardial cavity was powdered with talc in 3 cases; 10 patients underwent ligation and suture of the left coronary ostium. An aortocoronary bypass was performed in two cases with an autovenous graft and pulmonary arterial wall. Study of the early and late results of the operations revealed that ligation and suture of the left coronary ostium may be the method of choice in managing patients with origin of the left coronary artery from the pulmonary artery.

INTRODUCTION

Anomalous origin of one of the coronary arteries from the pulmonary trunk, as first described anatomically by Abbott in 1908 (1) and Abrikossof in 1911 (2), and clinically by Bland, White, and Garland in 1933 (3) is a rare cardiac defect. Nonetheless, because of the high mortality in infancy (4) and the possibilities of early diagnosis and surgical repair, it still attracts attention among clinicians. Pathological, hemodynamic, and surgical problems have been described extensively in the literature in recent years (5-10). These problems were also discussed at the first and third USA-USSR joint symposia in 1973 (11) and 1977 (12).

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At present, the main problems concerning the clinical signs, hemodynamics, and surgical approaches have essentially been solved by the studies of Edwards (13), Askenazi and Nadas (14), and Sabiston (15). The choice of surgical management procedures is one of the unsolved problems.

This paper presents an analysis of our experience with surgical repair of anomalous origin of the coronary arteries from the pulmonary trunk at the Bakulev Institute of Cardiovascular Surgery of the USSR Academy of Medical Sciences in the years 1966 through 1982.

MATERIALS AND METHODS

Twenty-two patients with anomalous origin of one of the coronary arteries from the pulmonary trunk were followed for 16 years. Twenty patients had anomalous origin of the left coronary artery and two had anomalous origin of the right coronary artery. Seven of the patients were under 1 year of age, and four were under 2 years of age. One patient was first examined at the age of 39 years. In our series, 10 of the patients were males and 12 were females (table 1).

TABLE 1. Distribution of Patients With Origin of Coronary Arteries From the Pulmonary Artery According to Sex and Age

	Age (years)								
Sex	0-1	1-2	2-3	3-5	5-10	10-20	20-30	30-40	Total
M	4	2	1	2	-	-	-	1	10
F	3	2	4	-	2	1	-	-	12
Total	7	4	5	2	2	1	-	1	22

In addition to routine clinical examinations, all patients underwent catheterization and angiocardiography, including left ventriculography, aortography, and selective coronarography from the right coronary ostium.

Five patients presented with a large aneurysm of the left ventricle. One 10-month-old baby with anomalous origin of the right coronary artery had been operated on for an interventricular septal defect, and an anomalous coronary artery was found during operation. In another 5-year-old patient, origin of the right coronary artery from the pulmonary artery was combined with an aortopulmonary septal defect. Signs of a left ventricular myocardial infarction were revealed in 3 patients, and signs of an associated mitral insufficiency were seen in 12 (table 2).

TABLE 2. Associated Defects

Main diagnosis	Associated defect	Cases
Origin of right coronary artery from pulmonary artery	Interventricular septal defect	1
	Aortopulmonary septal defect	1
Origin of left coronary artery from pulmonary artery	Ventricular aneurysm	5
	Mitral insufficiency	12
	Infarction	3
	Fibroelastosis	7

Surgery was performed on 16 patients. Three types of operations, reflecting the development of surgery at the time, were used (table 3). During the first few years, pericardiotomy and talcing of the pericardial cavity were used in an effort to develop collaterals. Subsequently, beginning in 1972, the anomalous artery was ligated or its ostium sutured from the lumen of the pulmonary trunk under conditions of artificial circulation. In recent years, three operations have been performed with restoration of the anterograde blood flow--in one case, along the left coronary artery by means of an autovenous graft, and in another, along the right coronary artery by reimplantation of its ostium into the ascending aorta with simultaneous reconstruction of the interventricular septal defect. One child underwent a transpulmonary aortopulmonary bypass graft.

In one case we performed patch closure of the aortopulmonary septal defect with interposition of the right coronary artery ostium. Nine patients were followed up from 1 to 7 years (mean, 4.2 years) after operation. Their results were evaluated by comparing the electrocardiographic (ECG) data from standard and thoracic leads. We also recorded and compared signs of myocardial ischemia (ST-segment shift) and left ventricular hypertrophy (size of the R wave in thoracic and monopolar leads). The change in heart size was calculated by the cardiothoracic index and Moore's index (16).

In order to evaluate the hemodynamics and contractility of the myocardium, we determined the ejection fraction of the left ventricle by the radiosotope method. The study was performed with a Picker Dynacamera-2 gamma camera. Radioactivity from the precardiac region was recorded on magnetic tape. The radiopharmaceutical used was ^{99m}technetium-labeled albumin (TCK-2) in the amount of 3.7^{108}-7.4^{108} Bq in a volume up to 0.5 ml. The data obtained were compared with preoperative data.

Exercise tolerance was evaluated on a bicycle ergometer with simultaneous ECG recording. Depending on body weight, the physical load was varied from 100 to 570 kg·m/min during the test. The following parameters were recorded: heart rate, respiratory rate, and arterial pressure. The loads were gradually increased in 25-50 W

increments, holding each increment for 3 to 6 minutes to maintain a steady state. The data obtained by comparing the indices were treated statistically with calculation of the numerical mean (M), mean error per group ($\pm$ m), and mean square deviation (σ). The significance was evaluated based on Student's tables.

TABLE 3. Operations

Pathology	Operation	Year	Cases
Origin of LCA from PA	Talcing of the pericardium	1966-1972	3
	LCA ligation	1972-1981	8
	Suture of LCA ostium under AC, resection of aneurysm	1980, 1980	2*
	ACB	1981-1983	2
Origin of RCA from PA	Reimplantation of the ostium into the aorta	1979	1
	Patch closure of intraventricular septal defect		
	Patch closure of APSD, interposition of RCA ostium	1981	1
Total			17

LCA = Left coronary artery. PA = Pulmonary artery. ACB = Aortocoronary bypass. AC = Artificial circulation. RCA = Right coronary artery. APSD = Aortopulmonary septal defect.

*In one case 6 years after pericardial talcing, an ACB was attempted; however, total obliteration of the pericardial cavity limited the operation to suture of the arterial ostium.

Immediate Results

Of 16 patients, 4 died in the early postoperative period (25 percent mortality). It must be noted that three of the patients died during the first few years of followup (table 4). Death was caused by acute heart failure in three cases and pulmonary complications in one (table 5). All of these patients died in infancy.

TABLE 4. Immediate Results

Pathology	Operation	Year	Age of patients	Survived	Died
Origin of LCA from PA	Pericardial talcing	1966-1972	6 mo, 1 yr 6 mo	2	1
	LCA ligation	1972-1981	4.5 mo, 2 yr 9 mo	5	2
	LCA suture aneurysmectomy	1980, 1980	7.5 yr, 9 yr	2	-
	ACB	1981, 1983	1 yr 4 mo, 10 yr	1	1
Origin of RCA from PA	RCA ostium interposition	1979	10 mo	1	-
	RCA ostium interposition, patch closure of APSD	1981		1	-
Total				12	4

See table 3 for abbreviations.

TABLE 5. Causes of Death in Patients With Origin of the Left Coronary Artery From the Pulmonary Artery in the Immediate Postoperative Period

Patient	Age	Operation	Cause of death
Ch.	6 mo	Talcing	Acute cardiac failure
S.	2 mo	LCA ligation	Acute cardiac failure
P.	8 mo	LCA ligation	Pulmonary complications
F.	1 yr 2 mo	ACB	Acute cardiac failure

See table 3 for abbreviations.

Late Results

In the remote period, one patient died 4 months after ligation of the left coronary artery. This was a critically ill infant with poorly developed intercoronary collaterals. In spite of the presence of an anastomosis to the pulmonary artery on the angiocardiogram, indicating a shunt of blood, the operation did not yield the desired result. Attacks of pain and heart failure persisted in the infant. Because of the extremely critical condition and the presence of acute disturbances in myocardial contractility (the preoperative ejection fraction was 0.23), it was impossible to perform another bypass operation (table 6). At autopsy the myocardia of both ventricles were heavily damaged by scars and collaterals were virtually absent.

TABLE 6. Cardiac Catheterization Data in a Patient Who Died 2 Months After Ligation of the Left Coronary Artery

Data	Value
Left ventricle pressure (mm Hg)	74/8 ± 18
Aortic pressure (mm Hg)	75/45 (mean, 60)
Ejection fraction of left ventricle	0.23

Eight of the patients were examined 1 to 7 years after operation. Of the two patients surviving pericardial talcing, an aortocoronary bypass was attempted in one, but because of technical difficulties (adhesions, pericardial obliteration), we had to suture the arterial ostium. Complaints and poor results were seen in one female 7 years after undergoing pericardial talcing. Signs of severe hypertrophy of the left ventricle and left atrium were seen in the ECG. We found an increase in the RV_{5-6} waves to 27-30 mm; the sum of $RV_5 + SV_1$ averaged 39.5 mm; the ST segment in leads I, AVL, and V_{5-6} was elevated by 4-5 mm.

X-ray revealed pulmonary hypervolemia, manifested as an increase in the caliber of the pulmonary arterial vessels. Moore's index was increased from 50-58 percent. The cardiothoracic index was 71 percent. An enlargement of the left atrium and severe dilatation of the left ventricular cavity with an elongated arch were seen in the anteroposterior and oblique projections. Cardiac catheterization and angiocardiography revealed an elevated end-diastolic pressure in the left ventricle (19 mm Hg) with dyskinesia of the anterolateral wall. Selective coronarography showed a small number of intercoronary anastomoses. The left coronary artery and pulmonary arterial trunk were slightly opacified. The patient refused surgery and died 1 year after examination.

All patients undergoing ligation and suture of the arterial ostium, as well as left coronary bypass, felt well and exhibited normal development.

Comparison of the pre- and postoperative ECG data for all patients showed normalization or a statistically significant decrease in left ventricular hypertrophy (table 7). The RV_{5-6} waves and the $RV_5 + SV_1$ sum was 10.6 ± 5.0 mm and 18.4 ± 9.7 mm, respectively. The AVL leads also showed a statistically significant decrease to 7.9 ± 4.9 mm, and SV_{2-3} fell from 19.0 ± 3.4 mm to 9.7 ± 4.7 mm. It must be noted that the marked decrease in left ventricular hypertrophy was seen in two patients with a relatively short observation time (1-2 years). Signs of left ventricular hypertrophy were absent in six patients 4-6 years after operation (figure 1).

In the right thoracic leads (V_{1-2}), normal P waves were observed with a statistically significant reduction in their duration to normal levels (0.07 ± 0.01 sec), indicating no hypertrophy of the left atrium (figure 2, table 7). In no cases did we note any shift in the ST segment (figure 3, table 7).

TABLE 7. Dynamics of ECG Indices in the Late Postoperative Period

Type of operation	Before operation: LVH $RV_{5-6} \geq 26$ mm	$RV_5 + SV_1 \geq 35$ mm	$R_{AVL} \geq 11$ mm	$SV_{2-3} \geq 10.5$ mm	Before operation: LAH P-wave duration (sec)	Before operation: Ischemia ST-segment shift (mm)	After operation: LVH $RV_{5-6} \geq 26$ mm	$RV_5 + SV_1 \geq 35$ mm	$R_{AVL} \geq 11$ mm	$SV_{2-3} \geq 10.5$ mm	After operation: LAH P-wave duration (sec)	After operation: Ischemia ST-segment shift (mm)
Ligation and suture of LCA ostium	10	30	29	20	0.12	4	9	9	4	6	0.06	--
	26	35	7	16	0.12	3	6	12	3	7	0.08	--
	27	52	24	25	0.12	5	8	26	16	18	0.07	--
	29	36	17	15	0.12	3	15	30	10	14	0.08	--
	24	39	15	20	----	5	8	10	4	5	0.09	--
	23	45	22	17	0.13	5	20	30	12	10	0.08	--
ACB	30	50	25	20	----	4	8	12	6	8	0.06	--
M ± m	24.1 ± 6.7	41.0 ± 12.5	19.9 ± 7.4	19.0 ± 3.4	0.12 ± 0.04	4.14 ± 0.9	10.6 ± 5.0	18.4 ± 9.7	7.9 ± 4.9	9.7 ± 4.7	0.07 ± 0.01	
σ	2.5	4.7	2.8	1.3	0.02	1.2	1.9	3.7	1.9	1.8	0.04	
p							< 0.05	< 0.05	< 0.05	< 0.05	< 0.05	< 0.05

LVH = Left ventricular hypertrophy. LAH = Left atrial hypertrophy. LCA = Left coronary artery.