Progress of Congenital Heart Disease: The Team Approach as It Includes the Anesthesiologist*

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Historical Background. No branch of medicine has progressed more rapidly and dramatically in the last 35 years than Pediatric Cardiology. Harvey's description of the circulation in 1628 (Harvey, 1628) and Leonardus Botallus' (Noback and Rehman, 1941) report on the fetal circulation were milestones in clarifying our understanding. Isolated clinical and pathologic descriptions of specific lesions written with amazing accuracy appeared in the 17th and 18th centuries. Sandifort (Sandifort, 1777) in 1777 presented details of a clinical examination correlated with necropsy findings in a cyanotic 12-year-old boy with pulmonic stenosis and a ventricular septal defect long before Dr. Fallot's famous report (Fallot, 1888). Chauveau and Marey (Chauveau and Marey, 1863) performed a right heart catheterization in 1861, but the technique was forgotten until Walter Forssman (Forssman, 1929), a urologist, reviewed it in 1929. Forssman even attempted an angiocardiogram, but the contrast injected was too dilute to be visualized and he was told by his associates that his work qualified "for a circus not a clinic." Despite such criticism, with Cournand and Richards, Forssman received the Nobel Prize in 1956.

The modern era of Pediatric Cardiology began with Dr. Maude Abbott, whose meticulous Atlas, published in 1936, presented precise descriptions of 1000 cases of congenital heart disease which she had personally examined from a pathologic standpoint (Abbott, 1936). She added a review of the development and comparative anatomy of reptilian, amphibian, and mammalian hearts. After reviewing the world literature related to the pathology of congenital cardiac malformations, she added her cases and published many comprehensive reports of individual lesions. As a result of her work, a few clinicians began to diagnose specific congenital heart lesions, although usually therapy was hopeless. Suddenly in 1938, the modern era of cardiac surgery began when Dr. Gross first successfully ligated a patent ductus arteriosus (Gross and Hubbard, 1939). Munro of Boston had suggested ligating a patent ductus as early as 1907 (Munro, 1907), but it was 30 years later before Strieder (Graybiel, *et al.*, 1938) attempted the technique. While Crafoord (Crafoord, 1965) in Sweden was ligating a patent ductus, the suture severed the vessel. It was necessary to clamp the aorta for 28 minutes. To his surprise, the patient did well and he recognized that this occlusion could be tolerated both by the heart and nervous system. Soon thereafter, in 1944, he successfully repaired a coarctation of the aorta in an 11-year-old boy (Crafoord and Nylin, 1945).

Successful surgery was a great stimulus to the diagnosis of acyanotic congential heart disease. About this time, Dr. Park, Professor of Pediatrics at Johns Hopkins, assigned Dr. Helen Taussig to the cardiology clinic suggesting that she accumulate data and correlate clinical information and pathologic findings on children with congenital heart disease. She quickly noted the repetitive patterns of developmental malformations and learned to make amazingly accurate diagnoses. Her monograph, published in 1947, was the first textbook on congenital heart disease (Taussig, 1947). Dr. Alfred Blalock recalled a small conference where he was discussing experimental production of pulmonary hypertension in animals using a systemic artery to pulmonary artery anastomosis. Dr. Taussig asked if he could increase the pulmonary blood flow in patients with pulmonic stenosis in the same way (Blalock, 1966).

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She was thinking of the many cyanotic children with decreased pulmonary blood flow as observed in tetralogy of Fallot or pulmonary atresia. No one knew the answer to three major questions: (1) Could a blue child tolerate anesthesia? (2) Could one occlude a pulmonary artery temporarily, especially in a blue child? (3) Would the arm tolerate ligation of the subclavian artery? Many animal experiments were carried out to attempt to answer these questions before they recognized that the answer to each was yes. The first Blalock-Taussig shunt for a tetralogy of Fallot was created successfully in 1944 (Blalock and Taussig, 1945). Modifications of the shunt procedure (Potts, *et al.*, 1946; Waterston, 1962) and Sir Russell Brock's (Brock, 1948) alleviation of valvular pulmonic stenosis followed quickly. Scott (Scott, 1955) at Vanderbilt in 1954 achieved open correction of a tetralogy with hypothermia, but it remained for Lillehei (Lillehei, Cohen, *et al.*, 1955) and Varco in Minneapolis, utilizing cross circulation, to first close a ventricular septal defect and completely remove right ventricular obstruction in 1955.

Gibbon had been working on a heart lung machine since the early 1930's and performed the first successful operation closing an atrial septal defect by this method in May, 1953 (Gibbon, 1954) Crafoord visited Gibbon in the late 1930's and his group simultaneously worked on a heart lung machine and developed a disc oxygenator. The advent of open heart surgery made possible accurate correction of the many complex congenital heart defects. Dr. Crafoord (Crafoord, 1965) considers the development of the Engstrom-Bjork respirator for anesthesia and postoperative ventilation a milestone in operative and postoperative care.

Developments in the physiology laboratory, including refinements in the technique of cardiac catheterization and particularly the addition of angiocardiography, as introduced by Robb and Steinberg (Robb and Steinberg, 1938), made precision in diagnosis possible. Improvements in the toxicity of contrast media made cineangiography less dangerous for small infants. Improved electronic equipment for recording and monitoring pressures and utilization of colorometric methods for rapid determination of oxygen saturations and indicator dilution increased the safety and efficiency of diagnostic cardiac catheterization. Rapid measurement of blood gases and electrolytes added another safety feature.

Transposition of the great arteries was first palliated in infancy in 1940 by Blalock and Hanlon (Hanlon and Blalock, 1948), by creating an atrial septal defect. This gave only temporary and moderate relief, but the frequency of this defect and its lethal characteristics were a continuing challenge to devise corrective surgery. Baffes (Baffes, 1956) attempted to reverse venous return. Senning (Senning, 1959) reported the first correction by plastic revision of the atria to reverse venous inflow in 1959. Mustard's (Mustard, 1964) use of an intra-atrial pericardial patch for total correction has vastly improved the prognosis in this previously hopeless lesion. In 1966, Rashkind and Miller (Rashkind and Miller, 1966) described the catheterization balloon technique for creating a large atrial septal defect, permitting survival through the critical neonatal period.

The achievement of success in correcting a complete truncus arteriosus or a complete atrio-ventricular canal captured the imagination of Rastelli and, based on a vast knowledge of anatomy, embryology, and physiology, has been achieved (Rastelli, Weidman, and Kirkland, 1965; Rastelli, Titus, and McGoon, 1967). The eight major congenital heart lesions now have excellent corrective procedures with acceptable mortality rates, which have diminished as preoperative diagnosis, surgical skill, and postoperative intensive care have improved. Rarer lesions are more hazardous and difficult to correct, especially in the younger child. The neonate, whose complex varieties of heart disease lead to cardiac failure and cyanosis early, needs early, aggressive diagnosis to avoid a 75% mortality. With ideal treatment, about ½ of the deaths can be prevented.

Auxillary aids in the care of a child with congenital heart disease lie in the fields of anesthesiology, hematology, biochemistry, and electronics. Landsteiner's (Landsteiner, 1901) description of blood groupings was essential for transfusions. A purified heparin, which allowed standardization and control of coagulation of blood, was a prerequisite to modern cardiac surgery. Pacemakers, defibrillators, prosthetic valves, and patches, now used routinely, have all been developed in the last few years.

The Medical College of Virginia assumed a position of leadership in modern aspects of cardiovascular and thoracic surgery under the direction of Dr. I. A. Bigger. There have been times, especially as open heart surgery began in the 1950's when this was a difficult role to maintain. The success of the

surgical effort is dramatically illustrated by the analysis of mortality statistics from the University of Minnesota for all open heart procedures on patients with congenital heart disease (Lillehei, Varco, *et al.*, 1967)

Progress of Open Heart Surgery for Congenital Heart Disease University of Minnesota

	Number of Cases	Operative Mortality
1954–1957	386	36%
1958–1962	986	22%
1963–1966	419	12%

At the Medical College of Virginia, my colleagues[1] have participated with me in a recent study to determine the incidence, morbidity, and mortality of surgically corrected congenital heart lesions over a six-year period. Our findings are stated below.

Materials and Methods. All patients, private or staff, who had surgery for correction or palliation of a congenital cardiac defect from birth to 20 years of age at the Medical College of Virginia Hospitals between January 1, 1966 and January 1, 1972 are included in this report. This six-year interval presents a complete evaluation of our recent surgical results. Every patient had a complete cardiac evaluation, including a history, physical examination, chest roentgenogram and electrocardiogram. Cardiac catheterization and angiocardiography were done except in children with a patent ductus arteriosus in whom these diagnostic procedures were frequently omitted. Decisions for surgery were made at a weekly conference in which the pediatric cardiologists, physiologists, radiologists, pathologists, and surgeons participated. Surgical procedures were divided approximately equally between the two surgical teams. During these six years, there were:

11,831	pediatric cardiology out-patient visits
2,985	hospital patients
557	children undergoing surgery for congenital cardiac defects
593	total surgical procedures for congenital cardiac lesions
45	deaths

[1] Dr. H. Page Mauck, Jr., Dr. Jon B. Tingelstad, Dr. Louise W. Robertson, and our surgeons, Dr. Richard R. Lower and Dr. Lewis H. Bosher, Jr.

Results. Five hundred fifty-seven children had major surgical procedures with the distribution as shown in Table I. The mortality for the common defects will be considered individually. Any death within the first month postoperatively is considered as surgical mortality. For the 593 procedures, there was a 7.58% mortality.

Atrial Septal Defects. Even a small atrial septal defect makes a preschool child a candidate for surgery. This was the most frequently corrected left to right shunt and was seen in 98 patients divided into the following types:

Secundum	72
Primum	15
With Partial Anomalous Pulmonary Venous Return	9
Single Atrium	2

All were catheterized and patients with a pulmonic to systemic flow ratio greater than 1.4:1 were referred for surgery at an appropriate age. The span was 11 months to 20 years. One infant with heart failure was corrected at 11 months. The mean age for correction was 6 years. There were no deaths in this group.

Ventricular Septal Defects. Eighty patients had closure of a moderately large ventricular septal defect which was the major lesion. The types are as noted (Table II) and include two with more than one defect and five with a LV-RA shunt. There were many associated defects, including atrial septal defects, aortic insufficiency, and patent ducti. Seven infants with intractable cardiac failure had pulmonary artery banding to decrease pulmonary blood flow caused by a large left to right shunt through the defect. Seven children required removal of a previously placed pulmonary artery band, in addition to closure of the defect. Two children have been reoperated for closure of a residual shunt. Severe aortic insufficiency from a redundant aortic cusp led to one surgical death. Of the seven infants under one year of age who required pulmonary artery banding, only four have good surgical palliation. Associated intra and extra cardiac defects complicated their course.

Patent Ductus Arteriosus. A patent ductus arteriosus has been interrupted in 90 patients between the ages of 3 months and 19 years. Almost all have been seen postoperatively and discharged as

TABLE I
INCIDENCE CONGENITAL DEFECTS

Defect		Patients (557 Patients) (593 Procedures)	Expired (45)	MCV Mortality % (8.07%) (7.58)
Acyanotic				
Atrial Septal Defect		98	0	0
Ventricular Septal Defect		87	4	4.6
Patent Ductus Arteriosus		90	2	2.2
Coarctation		34	1	2.9
Pulmonic Stenosis		36	0	0
Aortic Stenosis		15	0	0
Vascular Rings		5	1	20
Miscellaneous Acyanotic		3	0	
Cyanotic				
Tetralogy of Fallot		118		
Shunts	61		8	13.1
Open Correction	63		3	4.7
Thoracotomy	1		0	0
Pacemaker	1		0	0
Transposition of Great Vessels		21		14.2
Palliative	17		3	17.6%
Mustard Procedure	4		0	0
Rare Cyanotic Lesions		50	23	46

cured. Two children, one an infant, died of technical problems during surgery. Because of cardiac failure and marked cardiomegaly, eleven infants were surgically corrected under one year of age. Ideally, surgery is deferred until the child is greater than 2 years of age. Several premature infants on whom the diagnosis was confirmed had spontaneous closure of their ductus.

Coarctation of the Aorta. Coarctation of the aorta was corrected in 34 patients at a mean age of 6 years. All patients had preoperative aortography. Four patients required surgery in infancy for intractable congestive failure, of whom one infant died. Successful correction of a complete interruption of the aortic arch has been done on two occasions.

TABLE II
VENTRICULAR SEPTAL DEFECTS (87)

Types:			
Corrected		80	
Single	73		
(Removal of Bands 7)			
Multiple	2		
LV-RA Shunts	5		
Banded Pulmonary Artery		7	
Ages:			6 Weeks–20 Years
Deaths:			4 (4.6%)
Correction	2	(2.5%)	
Pulmonary Artery Bands	2	(28.5%)	

Pulmonic Stenosis. Pulmonic stenosis is commonly associated with other defects, but was seen as the primary problem in 36 children who had an intact ventricular septum and a right ventricular to pulmonary artery systolic gradient between 50 and 160 mm. Hg. The age span was 11 days to 18 years and two infants were less than 1 year of age. The types are as follows:

Valvular	28
Supravalvular	1
Primary Infundibular	4
Rubella with Valvular Pulmonic Stenosis and Peripheral Pulmonic Stenosis and Patent Ductus Arteriosus	3

Associated defects included atrial septal defect and peripheral pulmonic stenosis in five. An associated rubella syndrome with a patent ductus occurred in three babies. Secondary infundibular obstruction required surgical alleviation in about ½ of the valvular cases. All survived and were improved.

Aortic Stenosis. Although 80 cases of aortic

stenosis were studied in the cardiac catheterization laboratory in this six-year period, only 15 were referred for surgery. The age span was 3 years to 19 years. The types were as follows:

Valvular	10
Subvalvular Membrane	2
Idiopathic Hypertrophic Subaortic Stenosis	1
Supravalvular	1
With Severe Aortic Insufficiency	1

Criteria for operation were a gradient in excess of 60 mm Hg and electrocardiographic evidence of left ventricular hypertrophy and strain. This age group is older than for most other lesions and included no infants. Those with valvular stenosis underwent valvuloplasty; only one prosthetic valve was utilized in a 15-year-old girl with severe aortic insufficiency. A teflon patch was employed to widen the boy's supravalvular aortic stenosis. Four patients with aortic insufficiency initially continued to manifest mild valvular incompetence.

Vascular Rings. Five patients with an aortic vascular ring were repaired under 8 months of age. One infant with a left pulmonary artery arising from the right pulmonary artery causing a vascular sling did not survive the surgical procedure. All others had relief of their stridor and have improved after 1 to 5 years. One infant also had a shunt for severe tetralogy.

Miscellaneous Acyanotic Lesions. An anomalous right coronary artery arising from the pulmonary artery was repaired successfully by direct anastomosis to the aorta in a 12-year-old boy. An arterio venous fistula between the external carotid artery and the internal jugular vein was also repaired last year.

Tetralogy of Fallot. One hundred eighteen children had 126 primary surgical procedures for tetralogy of Fallot (Table III). In general, a Waterston's shunt (aorta to right pulmonary artery anastomosis) was the procedure of choice in the first three months of life, a Blalock-Taussig shunt (subclavian to pulmonary artery) being preferable if one was required in a slightly larger child. Thirty-six infants had shunts created within the first year of life, of whom twenty eight survived. In 1970, six infants with a complete pulmonary atresia with a ventricular septal defect had emergency surgery attempted in the first few weeks of life; only two survived. There were no deaths in children who had shunt procedures over 1 year of age. The mortality for the shunt, including those with pulmonary atresia, is 13.1%. Seven patients required revision of their shunt at some time postoperatively. The majority were treated with digitalis in the postoperative period, since cardiac failure was a frequent transient complication.

TABLE III
TETRALOGY OF FALLOT
(126) Primary Procedures
(118) Children

Types:			
Shunts			61
Open Correction			63
Previous Shunts		28	
No Previous Shunts		35	
Thoracotomy			1
Pacemaker			1
Secondary Procedures			11
Revision of Shunts	7		
Secondary Repairs	4		
Deaths:			
Mortality Shunts			8 (13.1%)
Mortality Correction			3 (4.7%)
Total			11 (8.8%)

Open correction of tetralogy in 63 patients was successful in 60, giving this a mortality of 4.7%. This surgical procedure is generally reserved for those over 40 pounds with an age distribution as noted in fig. 1. The youngest child was 3 years old and weighed 27 pounds. Of the 63 children undergoing open correction, 35 had no previous shunt. Twenty-eight had shunting surgery in the past and 8 had both procedures during the six years of study. Transient cardiac failure postoperatively occurs frequently, but responds well to digitalis and diuretics. Permanent complete heart block occurred in 2 children; one died suddenly soon after the operation; the other now has a pacemaker implanted. Four children have required reoperation for residual ventricular defects or inadequate relief of their pulmonary stenosis. All have been followed postoperatively and 10 recatheterized. Many have major extra cardiac lesions involving the brain, kidney, gastrointestinal tract, or skeleton.

Transposition of the Great Arteries. In many series, transposition has comprised up to 20% of necropsy cases in cyanotic congenital heart disease under 1 year of age. Twenty-one cases of trans-

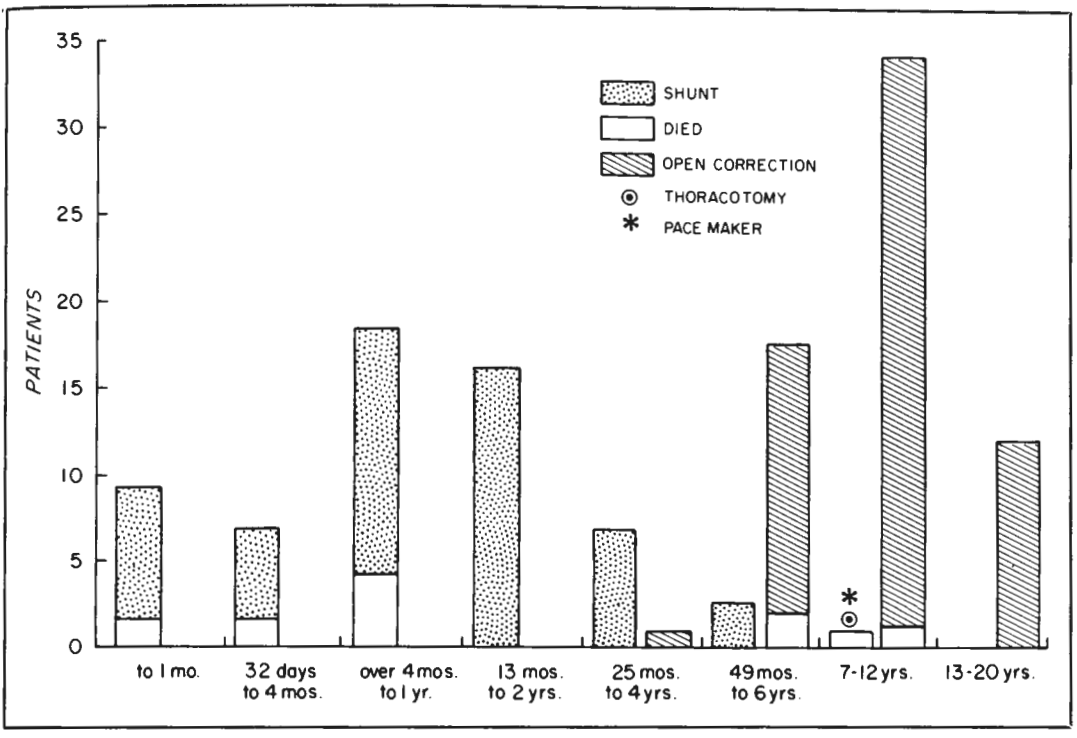

Fig. 1—Tetralogy of Fallot surgical therapy according to age.

position of the usual variety occurred in our series. Others associated with a single ventricle, bilocular heart, or tricuspid atresia are included with miscellaneous cyanotic lesions (Table IV).

All infants with transposition of the great arteries had palliative procedures in infancy, usually a balloon septostomy or a Blalock-Hanlon septectomy. In only two did the initial balloon septostomy produce adequate mixing. Usually the Blalock-Hanlon surgical procedure followed within a few weeks. Infants with associated ventricular septal defects required a concomitant pulmonary artery band and one with an associated ductus had it ligated. A palliative shunt was performed on two infants with associated pulmonic stenosis. The palliative procedures had a high risk. There were no deaths directly related to the balloon procedure. Four children have thus far had complete correction by the Mustard technique and improved postoperatively. One boy with residual pulmonary hypertension died this month, 33 months after the correction.

Miscellaneous Cyanotic Lesions. Fifty children with rarer cyanotic congenital defects were seen, of whom thirty-six were less than one year of age (Table V). Of the eleven cyanotic children with tricuspid atresia, nine were infants and two were older, with an age span from 1 day to 17 years.

TABLE IV
TRANSPOSITION OF THE GREAT VESSELS (21)

Procedures		Expired	Mortality %
Catheterization	11	0	0
Balloon			
Surgical			
Palliative	15		17.6%
Blalock–Hanlon 11		2	
Blalock–Hanlon and Banded Pulmonary Artery 2		1	
Shunts 2		0	
Correction		0	
Mustard 4			
Total Surgical Procedures	30		10%
Total Patients	21		14.2%

TABLE V
MISCELLANEOUS CYANOTIC LESIONS (50)

Defects		Patients	Expired
Tricuspid Atresia		11	1
Total Anomalous Pulmonary Venous Return		3	2
Single Ventricle		6	2
Banded	4		
Shunt with Pulmonic Stenosis	1		
Thoracotomy	1		
AV Canal		5	2
Splenic Syndromes		5	2
Multiple Mixed		20	14
Pulmonary Atresia	4		
Mitral Atresia	3		
Hypoplastic Right Heart	1		
Truncus	4		
Double Outlet Right Ventricle	2		
Ebstein's	3		
Hypoplastic Left Heart with Mitral Stenosis	2		
Multiple Defects	1		
Total		50	23

Nine required shunts to increase pulmonary blood flow, but two cases with unusually large pulmonary blood flow were palliated by banding the main pulmonary artery. The two older children had their second shunt created during the past year; one a 17-year-old high school senior has won a trip to Europe this summer on a singing tour.

The others are a discouraging group most of whom have been palliated by shunts or banding the pulmonary artery. A balloon septostomy did not significantly improve the condition of one child with pulmonary atresia. One atrio-ventricular canal was corrected and survived. A boy of 17 with a truncus arteriosus, Type I, had an attempted Rastelli procedure, but had irreversible pulmonary hypertension. The total mortality was 46%, but 36 were less than one year of age.

Conclusion. During the last six years, 557 children have had 593 surgical procedures for correction or palliation of their congenital heart defects with a mortality rate of 7.58%. One hundred fourteen patients were less than one year of age. Of the 368 acyanotic patients, 2.16% died at surgery or of related causes in the following month. Tetralogy of Fallot was the most favorable cyanotic lesion with a mortality for shunts in early childhood of 13% and for open correction of 63 patients it was 4.7%. Cooley's (Cooley and Hallman, 1966) recent review of his experience over a 10-year period reports the mortality for shunts as 7.2% and for open correction 15%. At Johns Hopkins University Hospital, 187 patients, ages 2 to 40 years, underwent complete correction of tetralogy in the last five years with a 9.6% mortality (Bender, *et al.*, 1970).

Our study showed 18 of 21 children with transposition of the great arteries survived. While other children with complex cyanotic lesions had a 46% mortality, this included many high-risk infants in whom an aggressive approach for palliation was justified because their prognosis was, otherwise, hopeless.

Advances in cardiac surgery have been great in the last 30 years, but further progress is anticipated, especially in infants. A cooperative effort involving referring physicians, pediatric cardiologists, physiologists, anesthesiologists, surgeons, and intensive care nurses is required to continue this trend.

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