16 PROGRESS IN CARDIOLOGY

Edited by

PAUL N. YU

and

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600 Washington Square
Philadelphia, PA 19106-4198
U.S.A.
(215) 922-1330

OTHER BOOKS IN THE SERIES

Progress in Cardiology 2—1973—\$15.00
Progress in Cardiology 4—1975—\$15.00
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Progress in Cardiology 12—1983—\$45.00
Progress in Cardiology 13—1985—\$30.00
Progress in Cardiology 14—1986—\$48.50
Progress in Cardiology 15—1987—\$35.00

The Library of Congress has catalogued this serial publication as follows:

Progress in cardiology. 1-Philadelphia, Lea & Febiger, 1972-

v. ill. 26 cm. annual. ISSN 0097-109X 1. Cardiology—Periodicals. RC681.A1P74 616.1'2'005

77---157474

ISBN 0-8121-0409-9 (v. 2)
ISBN 0-8121-0451-X (v. 3)
ISBN 0-8121-0509-5 (v. 4)
ISBN 0-8121-0578-8 (v. 5)
ISBN 0-8121-0596-6 (v. 6)
ISBN 0-8121-0621-0 (v. 7)
ISBN 0-8121-0621-0 (v. 7)
ISBN 0-8121-0621-0 (v. 9)
ISBN 0-8121-0728-4 (v. 9)
ISBN 0-8121-0815-9 (v. 10)
ISBN 0-8121-0815-9 (v. 11)
ISBN 0-8121-0911-2 (v. 12)
ISBN 0-8121-0911-2 (v. 12)
ISBN 0-8121-1018-8 (v. 14)
ISBN 0-8121-1018-8 (v. 14)
ISBN 0-8121-1014-7 (v. 15)
ISBN 0-8121-1144-3 (v. 16)

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Printed in the United States of America

PREFACE

Volume 16 of "Progress in Cardiology" marks the end of our active editorship.

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In 16 years, cardiology has advanced its frontiers dramatically. The first chapter of Volume 1 in 1972 was entitled "Computers in Cardiology" by J.P. Blackburn of the Westminster Hospital in London, England. Since then, we have seen the practical realization of signal-processing in a clinical context, such as on-line analysis of data in the catheterization laboratory. In contrast with this opening chapter in an expanding field, the final chapter was on a timeless theme; pulmonary edema by Dr. Grant Lee of Oxford.

In the intervening 15 years, we have seen the development of coronary artery bypass grafting from the early endeavors at the Cleveland Clinic in the late 1960's and early 1970's to the refinements of modern coronary artery surgery such as the use of the internal mammary artery as a graft for the artery with the most severe disease. Also in Volume 1, Favoloro presented an analysis of the results of 4,422 revascularization procedures carried out at the Clinic from 1962 to 1970, including internal mammary implants-now obsolete. When such data were presented at the VI World Congress of Cardiology in London in 1970, there was intense interest, but also, no little skepticism. Now we can see the fruits of the early struggles in the excellent long-term results of coronary artery bypass grafting and the striking improvement of symptoms in most patients. Naturally, some problems still remain, but many have been solved. Percutaneous coronary angioplasty was not an accepted form of treatment for coronary

occlusive disease in 1972; now it has a firm place in therapy, and each year, new and more effective catheters are developed, while laser techniques are under extensive investigation, and percutaneous balloon angioplasty is being used to treat valvar lesions.

Although thrombolysis for coronary artery occlusion was practiced in the Soviet Union in 1976, it was not widely accepted as a method of salvaging ischemic myocardium as it is now, even though many problems remain.

The exciting developments in noninvasive studies, especially the introduction of sector-scanning two-dimensional echocardiography and color doppler echocardiography has transformed diagnosis, making invasive investigation unnecessary in certain instances.

Increasing sophistication in nuclear cardiology has greatly improved the assessment of regional and global myocardial ischemia and function in circumstances of minimal invasion of the patient. Now the application of positron emission tomography, of computerized tomographic scanning, and magnetic resonance imaging to cardiac disease has advanced knowledge still further. Exciting new fields of research lie in the studies of the velocity of blood flow with MRI imaging, and the spectroscopic use of MRI to study the biochemistry of the myocardium. Perhaps the most exciting development of all is the application of molecular biology and genetics to cardiology, that will almost certainly be featured in future volumes of Progress in Cardiology.

The progress in techniques of endomyocardial biopsy and in its interpretation has enormously assisted in the dramatic results of cardiac transplantation and has helped to develop our understanding of the cardiomyopathies.

Volumes 1 through 16 of Progress in Cardiology have covered all the developments of cardiology, providing a balanced assessment by experts. The introduction in Volume 5 of periodic symposia on important current topics has proved most successful. Following the first symposium on coronary blood flow, Volume 6 was devoted entirely to the surgical aspects of coronary artery disease, so important and immediate was the subject considered to be. Volume 10 was devoted to predictions for the future, and dedicated to Sir John McMichael for his outstanding contributions to cardiology, beginning with his pioneer work in cardiac catheterization.

Looking back at cardiology over this century, it is apparent that the discipline has passed through several evolutionary phases: the development of electrocardiography; the precise measurement of pulse waves, phonocardiography; cardiac radiology, all heralded the emergence of cardiology as a precise clinical science. The dramatic advances made possible by cardiac catheterization and angiocardiography opened a window and flooded the field with light and understanding. Now it became possible to diagnose precisely the state of hemodynamics and functional disorders on the basis of physical signs at the bedside. The information gained by cardiac catheterization and angiocardiography provided the opportunity for which cardiac surgeons had long been waiting.

When the next major advance occurred, the heart-lung machine, they grasped eagerly the possibilities afforded for open heart surgery. As methods of diagnosis improved and better cardiovascular drugs became available, the range of therapy for cardiovascular disease expanded. Excellent echocardiographic equipment and techniques enabled precise diagnosis to be made, especially in neonatal patients, making angiography often unnecessary in this group. The apotheosis of expert team work, including so many different disciplines, and many specialties was manifest in cardiac transplantation.

Cardiologists today live in a fascinating era

of successful interventional therapies, such as angioplasty and valvarplasty, and also in an era of immensely sophisticated, complex and expensive investigation. It is of crucial importance that the basic clinical skills are not lost, but are used to guide the clinician investigator to the most appropriate investigations that lead to accurate diagnosis and the best possible treatment.

During the past 40 years, international relationships in cardiology have been developing steadily, culminating in the merger of the International Society of Cardiology with the International Cardiology Foundation in 1978. These relations have been greatly encouraged by the American Heart Association and the American College of Cardiology. Contacts in cardiology across the Atlantic between the U.K. and the Americas and Canada have always been strong, and it was with this in view, and mindful of the need for widespread co-operation in all parts of the world that "Progress in Cardiology" was begun at the suggestion of P.N.Y. We felt that it was important to have a regular publication with the widest possible geographic coverage and scientific and practical expertise. We envisaged chapters from a wide range of nations with differing cardiac problems appearing in each volume. The burgeoning of activity in Europe, the Middle and Far East, Africa and Oceana has further emphasized the need for a publication with a cardiologic message for all nations.

We are confident that, under the guidance of the new Editors: Dr. Douglas Zipes of Indianapolis and Dr. Derek Rowlands of Manchester, England, "Progress in Cardiology" will go from strength to strength. It will change, of course, to meet new challenges that lie ahead. We shall always be grateful to Mr. Kenneth Bussy and his immediate colleagues, and indeed to all at Lea & Febiger. We shall continue to take a lively interest in "Progress in Cardiology" as Consulting Editors, and wish the new Editors and all our readers every satisfaction.

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ACKNOWLEDGMENT

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The Editors and the Publisher wish to thank all those contributors past and present who helped to make PROGRESS IN CARDIOLOGY a successful enterprise. We particularly want to thank Dr. Alasdair Breckenridge, M.D., Professor of Clinical Pharmacology, The University of Liverpool, who was instrumental in obtaining many of the contributors to this volume.

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CONTRIBUTORS

The Editors and the Publisher wish to thank all those contributors had and present who televal to make PROCRESS IN CARDIOLOGY a successful encourse. We particularly want to thank

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HEART DISEASE IN THE ADOLESCENT

Jane Somerville, M.D., F.R.C.P.

Now that rheumatic fever has become a rare disease in many countries and congenital cardiac lesions are treated early in childhood, adolescents with cardiac problems have not merited much consideration.

Patients aged 12 to 19 years are considered adolescents. Where a service and interest is provided for adolescents, as in the National Heart Hospital, London, it is now clear that increasing numbers of patients require expert care as well as specialized investigative techniques, pacemaker services, and surgical skills. This is not because of a recrudescence of rheumatic diseases or an increased incidence of congenital cardiac anomalies which cause symptoms for the first time in the second decade. It is related to both residual and new problems appearing in children and infants who have already had surgical treatment for congenital heart disease. without which many would not have survived. This is a new medical community—the product of progress in the management of congenital heart disease. During the last 35 years, the survival and outlook for patients born with congenital heart disease has changed dramatically because of surgery, supported by accurate diagnostic methods, improved anesthesia and intensive care, safe cardiopulmonmary bypass and myocardial protection, new drugs, pacemakers and investigative techniques.

Besides cardiac problems, adolescents and young adults have many questions about contraception, pregnancy, employment, insurance, driving licenses, genetic risks and extracardiac

surgery. Law officers need guidance about the mitigating factors which the long-standing heart disease might justify. Expert advice must guide patient and authorities:

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The National Heart Hospital is a small specialist supraregional cardiac referral center with active surgical services since 1964. A service for children, not newborns, was established and has increased since 1968. Looking at numbers of In-Patient adult and adolescent admissions it is seen that by 1976 adolescent cardiac problems increased to provide 10% of the In-Patient work. In part this was due to interest in the subject and provision for adolescent needs. The majority of hospital admissions-about 93%-had congenital heart disease, the rest had acquired diseases, either primary rhythm disturbances. cardiomyopathy or myocarditis, or the effects of trauma. In 1985 new demands for beds for adolescent cardiac patients developed because increasing numbers of teenagers with complex congenital heart disease are now referred with terminal heart failure and pulmonary vascular disease to be assessed for cardiac or heart/lung transplantation.

Analysis of the reasons for admission of adolescent patients with congenital heart disease show that there are basically four:

- 1. First surgical treatment.
- 2. Medical cardiac admissions including investigation for 1.
- Re-operation in patients who have had radical repair or palliative procedures.

 Assessment for cardiac or heart lung transplantation.

It is appreciated that this experience is from a country where a well developed health service and cardiac surgery for infants and children have been established for more than 20 years. In other countries less fortunate, without such facilities, advanced congenital heart disease, rheumatic lesions (particularly mitral regurgitation and stenosis and triple valve disease) appear more frequently in the second decade.

FIRST SURGICAL PROCEDURES— ADOLESCENTS

Cyanotic Patients

Compared to 10 to 15 years ago, few patients with cyanotic problems have their first operation in the second decade of life. Some of the more complex anomalies which have not caused symptoms or severe cyanosis earlier needed help after the period of rapid growth (puberty). Such lesions associated with corrected transposition with pulmonary stenosis, Ebstein's anomaly with arrhythmias, double inlet ventricle or double outlet right ventricle previously balanced by mild pulmonary stenosis, mild tetralogy of Fallot and the rare case of transposition of the great arteries which survives with mild pulmonary stenosis and a large ventricular septal defect (VSD) appear in this category. Occasionally such lesions as common trunk, double outlet right ventricle and common atrioventricular canal and common atrium appear cyanosed with pulmonary hypertension and the decision about operability is difficult. Usually by this time, but not always, the pulmonary vascular disease is too advanced for a successful surgical outcome. However, the incidence of such problems depend on the available medical services. Where health services and special skills are readily available to all as in the U.K., these problems appear infrequently for the first procedure at the time of adolescence because they have had surgical treatment earlier.

Acyanotic Patients

Certain lesions become critical in adolescence or there are cardiac anomalies that must be repaired before a career can be embarked upon. Lesions which appear are shown in Table 1.

Sometimes large septal defects with hyper-kinetic pulmonary hypertension appear late and the degree of permanent pulmonary arteriolar damage may be difficult to assess in VSD, large persistent ductus arteriosus, aortopulmonary window and atrioventricular septal defects. Preliminary lung biopsy may be useful in determining the value and risks of surgical closure but must be viewed by an expert who understands. Infective endocarditis on VSD or left valve lesions can precipitate the need for cardiac surgery on otherwise mild lesions.

In centers with special expertise and interest in this group, the incidence of compensated complex lesions referred is rising in comparison with 10 years ago.

MEDICAL ADMISSIONS IN ADOLESCENTS

These were increasing steadily but with the advent of cross-sectional echocardiography there are fewer demands on In-Patient beds for

Table 1. Acyanotic Congenital Cardiac Anomalies
Appearing During Adolescence Needing
(Having) First Operation

ASD—Secundum, sinus venosus.

Atrioventricular septal defect
(Common atrium and ostium primum).

*VSD moderate/small

+ aortic regurgitation

+ pulmonary stenosis (moderate/mild) Pulmonary valve stenosis.

Absent pulmonary valve

(with VSD and infundibular stenosis)
Aortic stenosis—valvar

—subvalvar

-supravalvar

*Valve regurgitation.

*Mitral regurgitation—after ruptured chordae, prolapse (Coarctation of the aorta—rare now). Corrected transposition with VSD ± mitral regurgita-

tion.

Ebstein's anomaly with rhythm disorder.

invasive investigation and assessment after surgical procedures.

Admission to hospital is needed by adolescent "cardiacs" for:

- 1. rhythm disorders,
- 2. myocardial dysfunction,
- 3. endocarditis,
- 4. pulmonary hypertension.

About 70 to 75% have had surgical treatment in the first decade and are the late survivors of "modern" treatment for congenital and, less frequently, acquired heart disease.

Rhythm Disorders

Arrhythmias occur after radical reparative surgery in Fallot's tetralogy, transposition of the great arteries with intra-atrial baffles, tricuspid atresia with Fontan or modifications and shunts, atrioventricular defects, atrial septal defects and Ebstein's anomaly.

Problems of pacemaker failures, and changes are demanding. Although acquired heart block in relation to surgery is infrequent in Fallot's tetralogy and ventricular septal defect compared to 10 to 15 years ago, the incidence of heart block has increased with radical surgery for more complex anomalies such as corrected transposition where it is part of the natural history, septation for one ventricle hearts, transposition of the great arteries, Ebstein's anomaly and occasionally tricuspid atresia which poses difficulties for endocardial pacemaking. Sinus node disease with symptomatic bradycardias may occur late and unpredictably after open heart surgery despite normality in all routine investigations and signs. Thus, unexpected faints, particularly at celebrations, etc., should not be attributed automatically to vasovagal episodes or alcohol. Probably the damage was done during the surgery by snares, atrial incisions or ill placed retractors but in some there may be a congenital origin. Electrophysiologic studies have an important place in their understanding and management.

Repetitive pacemaker box changes are associated with complications particularly, and unfortunately, sepsis. It should not be regarded as a simple uncomplicated procedure without morbidity. Even mortality has occurred from septicemia and use of new untried models may have hazards. The patients are often short of access to veins by the time they reach adolescence so that epicardial pacing may need to be continued and thus fractured wires and electrical threshold problems with fibrosis may require thoracotomy. The technology of epicardial wires has not advanced as the development of better endocardial leads.

The management of complete heart block after Fontan's operation is difficult. Pacing from the coronary sinus is sometimes possible but it this fails, as it often does, epicardial pacing will be the only choice.

Our practice in the child (and sometimes the adolescent) has been to use epicardial pacing. particularly when heart block occurs at the time of surgery or the patient is at special risk of developing this, as in certain lesions such as atrioventricular defects with unusually long P-R interval, some examples of Ebsteins's disease. and corrected transposition. Permanent wires are placed at operation and subsequently the pacemaker is put in or behind the rectus sheath. Epicardial pacing has been associated with many problems, fractured wires, intraperitoneal migration of the box and premature failure. Where possible, endocardial pacing is instituted during adolescence if not before. Growth has not caused problems, probably because other factors interrupt good function before! Howeyer, when the adolescent is thin, the box may be disturbing. maignives to susmissioning sell-

Arrhythmias may precipitate heart failure—pulmonary congestion or right heart failure in patients with defects repaired particularly when the myocardium is already damaged, in complex anomalies already shunted or banded or as part of the natural history of the untreated lesion. Since their onset is often related to age, they can be expected to initiate problems in late adolescence. Particularly prone to this complication are repaired common atrium, transposition of the great arteries with intra-atrial baffles, and atrioventricular defects. Often the arrhythmias are complex, a combination of fast and slow requiring medication and pacemaking, or re-

sistant, needing multiple drugs in large and changing doses. The side effects of modern drug therapy can be many and serious, particularly with the routine use of amiodarone but it has been found to be most useful in control of ventricular tachycardia and supraventricular arrhythmias resistant to simple drugs.

Arrhythmias cause symptoms in previously compensated lesions demanding treatment for both. Where there is no surgical answer, intensive efforts to return the patient to sinus rhythm are vital. Too often efforts are made only to slow the ventricular rate in atrial fibrillation without attempts to revert it. The effectiveness of rate control with amiodarone and beta blocking agents has encouraged this disastrous policy which leads to deteriorating function and a downhill clinical course. Preservation of atrial contraction is important to most congenital lesions.

Ventricular arrhythmias in unoperated congenital heart disease are uncommon. However, ventricular tachycardia in tricuspid atresia is causing concern in these older patients. Ventricular tachycardia in tetralogy patients after operation and in other patients who have had an extensive right ventriculotomy (looking like an old infarct at necropsy) can be suppressed and there may be a case for surgical extirpation of the area if it can be identified. We still have the problem of unexpected sudden death in this group. Some of the "collapses" are in response to atrial flutter. Often, despite extensive routine testing, the patient who dies has not been identified as "at risk". Another related problem is the management of asymptomatic ventricular ectopics which are frequent in these patients. If symptomatic or seen on routine electrocardiography. Holter monitoring is vital both as an indicator of severity, and frequency, and foci, and as a baseline for monitoring the effectiveness of therapy. Current policy is to treat those which do not disappear or increase on effort, if multifocal, and if in doublets or triplets.

Primary arrhythmias unassociated with structural congenital anomalies cause symptoms in the adolescent. The commonest, the supraventricular tachycardias with Wolff-Parkinson-White (WPW) or Lown-Gannon-Levine (LGL) variant syndrome often appear around or after puberty, causing angina, syncope and less frequently heart failure, interfering with school or university work at a time when maximum effort is needed to pass examinations. Drug therapy often may cause intolerable side effects for the active teenager and surgery or ablation and perhaps a complex pacemaker may offer the only solution to the resistant case. Ablation has hazards of rupture and pericardial tamponade. Ventricular tachycardias with myocardial disease with long Q-T syndromes, or with other life threatening ventricular arrhythmias are also seen.

The demand for expert electrophysiologic studies and pacemaking for the adolescent cardiac patient is clear and facilities should be available wherever these patients are managed.

Myocardial Dysfunction

Adolescent patients require hospital In-Patient treatment for cardiac failure, for arrhythmias with manifestations of myocardial dysfunction and for investigation of cardiomegaly and dysfunction from cardiomyopathy or unexplicable cardiomegaly after earlier reparative surgery for congenital heart disease. Cardiac problems due to hypertrophic cardiomyopathy or dilated (congestive), probably postmyocarditic, also appear at this age. Unfortunately. "iatrogenic" cardiomyopathy is appearing in patients treated earlier with adriamycin for leukemia, osteogenic sarcoma and other tumors. Now that cardiac transplantation has offered dramatic palliation, these patients have been referred more frequently over the last 2 years and make heavy demands on all facilities as they can appear in a terminal phase with the tumor "cured" and the heart irreversibly damaged by chemotherapy or chest radiation.

An unfortunate cause of late heart failure is intraoperative damage to the myocardium during open heart surgery 10 to 15 years earlier. Patients present with minimal residual lesions, valvar regurgitation or stenosis, or small defects which cannot account for the gross inyocardial dysfunction. This state is caused by prolonged ischemia from lack of effective invocardial pro-

tection and inadequate coronary perfusion during cardiopulmonary bypass. It may be useful to study the operation report to find the cause of such damage although details of ischemic time, cross clamping the aorta, and coronary perfusion are often missing. The electrocardiogram may suggest myocardial infarction dating from the time of surgery, or prolonged unexplained failure after the operation. Myocardial damage may manifest after repeated intracardiac operations to close a VSD, or repeated aortic valve surgery which predisposes to irreversible myocardial damage. Tricuspid and/or mitral regurgitation are also frequent complications, making the basic cause of the myocardial failure difficult to evaluate. The clinical features of serious myocardial dysfunction show 1 to 5 years later. At necropsy the heart appears dilated with large areas of fibrosis, sometimes with thickened fibroelastic endocardium lining the left ventricle resembling fibroelastosis-a probable consequence of subendocardial ischemia.

However good is the myocardial protection, some damage is inevitable during cardiopul-monary bypass. This increases the risks of third, fourth and fifth reoperations, and must be taken into account when one is timing repeated surgery.

Fortunately, serious myocardial damage following good surgery is now infrequent with new techniques for myocardial protection and more effective coronary perfusion. Some late myocardial dysfunction can be attributed to irreparable myocardial damage before operation for left outflow tract obstruction, severe pulmonary stenosis or chronic aortic or mitral regurgitation.

Even slight loss of left ventricular function has disturbing consequences in Fontan type operations, even with most modern techniques. This is posing problems in such patients.

There is another group of patients with congenital heart disease, operated upon or not, who have myocardial problems which cannot be attributed either to surgery or to severity of the lesion. This is an important subgroup in which the myocardial abnormalities are due to excessive dysplastic muscle (as found in hypertrophic cardiomyopathy) in the ventricular septum, and left or right ventricles. It is regarded as part of

diffuse congenital cardiovascular disease and both conducting arteries and coronary arteries also may be pathologically abnormal at birth. Much of this disease remains after good surgical treatment and may manifest as unexplained cardiomegaly and hypertrophy, arrhythmias, heart failure, or unusual electrocardiographic findings. How much such disorders will disturb the patient requires to be shown, but such examples as pulmonary stenosis with large left ventricle, mild aortic valve stenosis and irregular hypertrophy of the left ventricle behaving like hypertrophic cardiomyopathy, atrial and ventricular septal defects with abnormal left ventricle and abnormal electrocardiograms are examples of this problem which many neither recognize nor understand. However, awareness of the concept that fetal damage and disorder to the heart is not limited to holes in the septa. malformed valves and disordered connections may help to solve some of the mysteries in the adolescent patients with treated congenital heart disease. a la municipalitation influentiaventa

Endocarditis

Infective endocarditis occurs in adolescence and is the commonest cause for a long stay in hospital. It occurs in unoperated patients and less frequently in those who have already had surgical treatment.

Endocarditis on aortic and mitral valve lesions, and in relation to fixed subaortic and supra-aortic stenosis produces the classic features but, in VSD, referral and diagnosis is often late. Practitioners too readily give antibiotics for the attack of "flu," pleurisy or bronchitis without considering the possibility of endocarditis in relationship to the VSD. It is surprising how often repeated courses of antibiotics are given indiscriminately to this group of patients who are unusually reluctant attenders to doctors since they are used to good health. It is exceptional to see an infected persistent ductus now in our Nationals, as most ducts have been ligated. Sometimes an adolescent with unoperated complex pulmonary atresia turns up with infection on the aortic valve, or less commonly, presents with hemoptysis from endocarditis on a collat-