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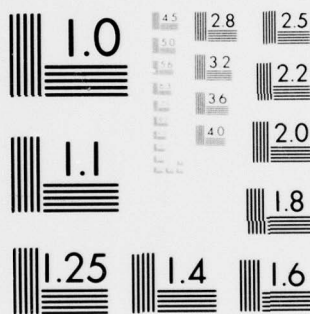
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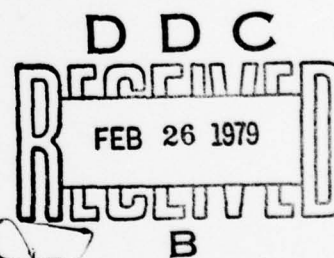
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PROGRESS IN CARDIOLOGY

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↙ Congenital heart disease patients are in many cases surviving into adult ages, presenting numerous new problems in management. Though anatomy may be corrected, physiology is not completely corrected. ↙

In coronary artery disease, the classic question persists: whether thrombosis precedes or follows myocardial infarction. Only 55% of a recent post-mortem series showed thrombosis, implying some other physiologic cause may underlie at least 45% of infarctions. Coronary spasm may occur in 100% of patients with unstable angina. Large clinical trials to limit infarct size are being begun in humans, following the demonstration that this is feasible in dogs, and that precordial mapping is a reliable measure of infarct size in humans.

Electrophysiologic techniques have dramatically increased our understanding of rhythm and conduction disturbances. The mechanism of producing and sustaining an arrhythmia can be worked out exactly in many patients with supraventricular tachycardia (SUT), ventricular tachycardia (VT) and Wolff-Parkinson-White (WPW) syndrome. Medical management of arrhythmias, however, still escapes a completely rational approach, in spite of numerous new drugs available. Electrophysiologic precision has also increased the opportunities for surgical treatment: VT and SVT with concealed bypass tracts may now be sometimes cured surgically, as well as the WPW syndrome.

Conquestive cardiomyopathy has been discovered to be associated with high titres of coxsackie virus. Anti-viral therapy may be of the future. Obstructive cardiomyopathy has been shown to have an abnormal but characteristic appearance on heart muscle biopsy. Sudden death remains the most important problem in this group, and high risk families have been identified.

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PROGRESS IN CARDIOLOGY

The attractive and balmy city of Nice, France, was the place for a recent meeting of some of the major lights in both American and European cardiology. From 15-19 May, professors from the US, England, France, Italy, and the Netherlands gathered to discuss the prevailing prominent dilemmas in cardiovascular disease in a conference sponsored by the University of Pennsylvania. The four daily sessions were devoted to, respectively, Valvular and Congenital Heart Disease, Coronary Disease, Electrophysiology, and Cardiomyopathies.

In addition to providing an opportunity to meet eminent cardiologists, the meeting presented the most recent data available on the major questions in heart disease. The only issue not discussed was that of the primary prevention (prevention before clinical illness develops) of arteriosclerotic heart disease. Though it is an area of a great deal of research, it seems to have little interest for cardiologists—perhaps because patients never come to see them until illness has developed.

Valvular and Congenital Heart Disease

Disease of the heart valves continues to be a very significant cause of morbidity and mortality in the overall spectrum of heart disease, though it has been surpassed in overall numbers by the epidemic of arteriosclerotic coronary artery disease. Whether due to congenital, rheumatic, degenerative, infectious, or ischemic processes, the effects may be devastating to the patient, usually causing early debility and early demise.

Prosthetic Heart Valves—The fundamental development that has transformed the outlook for these patients over the last two decades has been the use of prosthetic valves. Dr. Malcolm Silver (Univ. of Toronto, Canada) reviewed experience to date with the various kinds of valves, of which there are several types: ball valves housed in stainless steel cages, tilting discs, and tissue grafts. Unfortunately, their presence in the body may lead to a whole new set of problems and complicated clinical courses. The main problems are thrombosis, infection, hemolysis, and endocardial thickening due to turbulence. All metal and plastic valves represent a nidus for thrombosis to occur. Thus anticoagulants must be administered for the life of the patient, and being on these has its own risks.

The various tissue grafts cause less turbulence and hemolysis. Durability has been their main problem. Dead tissue in the body tends to calcify. Human allografts (transplanted human valve tissue) will calcify and begin to lose function in four to six years. *Fascia lata* grafts will do so in six to eight years. The most recent innovation has been the porcine heterograft (pig valve), which has been used for about five years, with substantially less complications than the previous

types to date. The main question now, so far unanswered, is how long will they last without being destroyed by calcification?

The present indications for and results of surgery for valvular disease were reviewed by Dr. Nathaniel Reichech (Univ. of Pennsylvania, Philadelphia). He summarized operative mortality at good medical centers as about 3-5% for single aortic valve replacement, 6-8% for single valve replacement, 8-12% for double valve replacement, and 6-9% for single replacement with coronary artery bypass graft. Closed commissurotomy carries a risk of 1-2%. In mitral valve surgery peri-operative mortality was correlated with the size of the left atrium—a larger atrium indicating increased operative risk.

The background against which surgical results must be compared is the natural history of valve disease in the unoperated state. Aortic stenosis is a particularly dismal situation, being recognized relatively late: 50% of these patients are dead two years after the diagnosis is made if surgery is not performed. Aortic insufficiency, mitral stenosis, and mitral insufficiency all have more prolonged survival curves which are similar to one another. One can identify subsets at higher risk: the presence of left ventricular hypertrophy and left atrial enlargement carry added risk.

The long term post-operative survival indicates that there is definite improvement in survival curves. For closed commissurotomy in mitral valve disease, the five-year mortality rate is 8-9%, the ten year 15%. For mitral valve replacement with the Starr-Edwards (ball valve) prosthesis, 50% survive 15 years, and the preponderance of deaths occurs in those with large left atria. The early results to date for the porcine heterograft look substantially better than for the Starr-Edwards. Causes of death in the patients after mitral valve replacement are diverse: congestive heart failure, emboli, hemorrhage, coronary arterial disease, arrhythmias, valve failure, and bacterial endocarditis.

Deaths after aortic valve replacement run 3-4% per year. Patients with a large left ventricle do much worse, as do those with coexistent coronary artery disease. Causes of death are usually congestive heart failure or arrhythmias.

The indications for valve replacement are still not exact, and remain one of the most difficult decisions in medicine. Reichech's recommendations are that a commissurotomy should be done if a patient has substantial symptoms due to mitral stenosis and if pulmonary artery systolic pressure is less than 80 mm Hg. A prosthesis should be inserted if the patient has shortness of breath on only mild exertion, and a pulmonary artery pressure greater than 80 mm Hg.

Mitral regurgitation requires replacement if there is severe regurgitation (3-4+ on pre-op catheterization) or if symptoms are class III, or if there is severe pulmonary hypertension. There is a tendency

to operate earlier in an attempt to save the failing left ventricle (overloaded by the extra blood volume), but proof is still lacking that this is beneficial.

For aortic stenosis, replacement is recommended for a gradient across the valve of greater than 75 mm Hg or an aortic valve area less than 0.75 cm^2 .

For aortic insufficiency, replacement is recommended for severe regurgitation (3-4+), for the presence of any symptoms, or for documentation of an enlarging left ventricle. There is some evidence that the use of vasodilators may be able to prolong the period of medical therapy prior to surgery.

Non-Invasive Diagnosis of Valvular Heart Disease—Dr. Harvey Feigenbaum (Indiana Univ. School of Medicine, Bloomington) spoke on developments in electrocardiography—a technique that is "non-invasive" because it does not involve the insertion of catheters into the vascular system. It is based on a sonar principle and produces a cross-sectional display of echoes created by a transducer placed on the chest wall. The display has certain characteristic features for various cardiac structures. Movement of the mitral valve, for instance, produces what is called the E-F slope. Variations in the E-F slope can be highly specific for the diagnosis of mitral stenosis: 98% specific in Feigenbaum's hands. Although the degree of severity of mitral stenosis cannot be determined from the E-F slope, suitability for commissurotomy can usually be determined.

The "floppy mitral valve" (or Barlow's) syndrome is a congenital syndrome in which there is minimal late mitral regurgitation. It is variously reported in from one to five percent of the population. Like other electrocardiographers, Feigenbaum finds it usually difficult to demonstrate, although one may be lucky and occasionally see its characteristic signs. The aortic valve is often difficult to see. Sometimes it may demonstrate thickening. The pulmonic valve is similar.

Electrocardiography is thus a very valuable tool for mitral stenosis, and can sometimes clarify other valve lesions. It requires, however, a high degree of skill in the electrocardiographer.

Congenital Heart Disease—The surgical correction of congenital heart disease means that patients with these lesions survive well into the adult age group. Dr. Alan Pearlman (Univ. of Pennsylvania, Philadelphia) described the present view of therapy for them.

An atrial-septal defect is the most common type, involving a communication between the right and left atria. Operative correction is simple, involving the placement of a patch. Even patients discovered in their forties can benefit from closure of the defect. On follow-up at one year, most patients had normal cardiac output at rest, but

it failed to rise normally with exercise. This residual abnormality has been interpreted as requiring lifelong follow-up, both to delineate the "natural" life cycle of the corrected abnormality and possibly to benefit the patient.

In ventricular septal defect, there is a good symptomatic relief and a decrease in pulmonary artery pressure, but the cardiac output in response to exercise is similarly blunted.

Tetralogy-of-Fallot exhibits similar response to exercise after correction, and there is a substantial incidence of conduction system defects after surgery. As there is a significant incidence of sudden death following correction as well, it has been attributed to this conduction system disease. Data were presented, however, strongly implying that sudden death is more closely correlated with the presence of ventricular ectopic hearts than with conduction system disease. This suggests a completely different approach to treatment to prevent this: that is, scrupulous suppression of ventricular ectopy rather than pacemaker therapy.

Coarctation of the aorta shows a definite decrease in mortality in operated patients. The hypertension seen in this disease may not be cured, however, and it may be progressive.

It is now clear that although anatomy may be corrected, physiology is not, and there is much yet to be learned about the long term course of these patients.

Coronary Disease

The coronary arteries are the first arteries which come off the aorta after it leaves the heart. They return to the heart muscle itself to supply it with oxygenated blood. It is arteriosclerotic obstruction of these arteries that leads to the vast majority of deaths from cardiac disease, via the mechanisms of myocardial infarction and conduction system disturbances.

Since myocardial infarction was first described in 1912, a debate has continued over the role of thrombosis in the development of the infarction itself. For many years it was thought that the precipitating event was the formation of clot in an artery—hence the diagnosis, coronary thrombosis, on millions of death certificates.

Dr. Malcolm Silver presented the results of his extensive autopsy series of coronary disease patients to bear on this question. He has found that the size of the infarct is not proportional to the degree of arteriosclerosis found in the coronaries. Arterial obstruction is firmly implicated, however, in that 95% of infarcts have at least 70% obstruction in the supplying artery. Occlusive thrombus is found in only 55% of patients. In Silver's view then, thrombosis is a secondary phenomenon following the infarct. This is the prevailing view at present. Similarly, in cases of sudden death, it is difficult to firmly demonstrate

even that an infarct occurred, as only 10% of cases show coagulation necrosis. Thus the major, Nobel prize-winning, 64 million dollar question in cardiology still remains: what causes a myocardial infarction? It is an enigma.

Exercise Stress testing—The only widely available noninvasive technique for the diagnosis of CAD is exercise electrocardiography. In this technique a patient is subjected to gradually increasing exercise while being monitored with multiple EKG electrodes. (Notwithstanding the obsession of the heart patient with his EKG tracing, the resting tracing is extremely poor for diagnosing coronary disease in the absence of a myocardial infarction.) The exercise test is also used to detect arrhythmias induced by exercise and to evaluate exercise tolerance. There is a good deal of ferment in the literature at present, and confusion among practitioners, about the validity of the test and the criteria for that validity. Dr. Joel Morgandoth (Univ. of Pennsylvania) reported his criteria based on comparison of exercise EKG results with coronary arteriograms. The results differ somewhat between symptomatic patients (with chest pain) and asymptomatic patients.

For symptomatic patients, there is a false negative rate of 20% and false positive rate of about 10% when the criterion for positivity is 1 mm of depression of the S-T segment on the tracing. Causes of false positives can be left ventricular hypertrophy, asymmetric septal hypertrophy, the mitral prolapse syndrome, or repolarization abnormalities due to drug therapy, conduction disturbances, or metabolic problems such as low potassium, or hyperventilation. It was found that the false positive rate can be reduced to 3% if 2 mm depression is used as the criterion for a positive test.

For asymptomatic subjects, the picture is much different. The predictive value when 1-mm depression is used is only 37-44%. Morgandoth's study found, however, that patients who genuinely had arterial obstruction on arteriography had an identifying characteristic—S-T depression usually lasting at least two minutes into the rest period after exercise, whereas patients with normal coronary arteries by arteriography would usually revert their S-T segments to normal by one-minute after exercise. If the appropriate but separate criteria for positive tests are used for symptomatic and nonsymptomatic patients, much of the confusion attendant upon the use of electrocardiography (which derives primarily from the presence of the "false positives") can be dispelled with these data.

Coronary spasm—One of the most intriguing dilemmas in modern cardiology has been the phenomenon of what is called variant angina, or Prinzmetal angina (after its describer). Its sufferers experience chest pain like that of patients with arteriosclerotic coronary disease, but get their pain at rest (atypically) or during sleep instead of on exertion, as do patients with conventional angina. Until recently, when it became clear that spasm may occur in coronary arteries, these people were told their pain was psychological in origin. Now it is possible

to diagnose this syndrome on the catheterization table, by inducing coronary spasm. Given ergot derivatives, spasm occurs and their pain is produced: Given nitroglycerin, the spasm is relieved and the pain disappears.

Dr. Attilio Maseri, (Milan, Italy) presented very provocative new data that illustrated the presence of coronary spasm in a class of patients in which it has hitherto not been known to occur. He studied by coronary arteriogram a group of patients with "unstable" (sometimes called "pre-infarction") angina. This is a syndrome in which a patient's chest pain occurs in an irregular and sometimes worsening pattern, often culminating in myocardial infarction. In 44 patients with this syndrome who underwent coronary arteriography, 100% of them showed coronary spasm during episodes of chest pain.

The implications of this finding are very startling. It appears that coronary spasm is not, as thought, a relatively rare phenomenon but probably occurs in all patients with unstable angina. Since many patients with chronic stable angina (of whom there are millions) may go into phases of unstable angina, there may be millions undergoing coronary spasm. Another startling process follows from the fact that most patients with myocardial infarction have prodromal periods of unstable angina before infarction. It may, in fact, be coronary spasm which underlies this prodrome and the infarction itself. Coronary spasm may be the key to the enigma of myocardial infarction.

Limiting infarct size—The present treatment of a myocardial infarct is limited to the prevention of complications—arrhythmias, cardiogenic shock, etc. The ultimate morbidity of a myocardial infarction depends on the degree of damage done to the heart muscle. Dr. Eugene Braunwald (Massachusetts General Hospital, Boston) described the theoretical foundation and present plans for research to limit infarct size after arrival at the hospital.

Data derived from experiments in dogs show that after coronary artery ligation, a detectable reduction in infarct size can be demonstrated up to six hours by the administration of drugs. There is no detectable effect at nine hours, however. Various drugs have been used. The most promising appear to be the beta-blocker propranolol and the enzyme hyaluronidase.

A fundamental requirement for a human study to limit infarct size is a noninvasive method to measure infarct size. It is presently thought that the technique of precordial mapping—whereby a phalanx of electrodes are placed across the chest and the summation of their perceived ischemic changes is calculated by a computer—can provide an accurate estimate.

A multicenter trial including 1500 patients is underway using precordial mapping to determine whether propranolol and hyaluronidase can reduce the damage of myocardial infarction.

Electrophysiology

The study of arrhythmias has advanced to the point that, through the use of cardiac catheterization in electrophysiology labs, it is possible to delineate in many cases the exact mechanism of producing and/or sustaining arrhythmia. Dr. Mark Josephson (Univ. of Pennsylvania) outlined the causes of supra-ventricular tachycardia in a series of 150 patients from his center. Sixty-seven percent are due to A-V nodal re-entry, 19% to the presence of a concealed bypass tract (WPW syndrome), 10% to sino-atrial or intra-atrial re-entry, and there is a small residuum—4 %—that are due to intrinsic automatism. Some of the cases of concealed bypass tracts do not have the usual EKG manifestations of the bypass tracts (deltawave). By studying these cases in the electrophysiology lab, it has been learned that such patients can be differentiated from those with A-V nodal re-entry by the presence of a retrograde P wave. The cases of A-V nodal re-entry have the P hidden within the QRS. The distinction between these types is important because the anti-arrhythmics (digitalis, propranolol, verapamil), given to patients with A-V nodal re-entry to slow A-V conduction, will make those with concealed bypass tracts worse. These patients must be given type I anti-arrhythmics (quinidine, Procainamide, or disopyramide).

Patients with ventricular tachycardia (VT) can also obtain benefit by being studied. The rhythm is induced by a technique called programmed electrical stimulation (PES). VT can also be initiated—or terminated—by a supra-ventricular beat. PES is successful, however, only in patients who have sustained VT, not in those with VT that is shortlasting or is caused by prolonged QT interval, or occurs on exercise. The mechanism has been found to be a circus movement about an area of delayed conduction. Class I anti-arrhythmics, by slowing the rate of conduction through the tract, are frequently very effective in interdicting this circus movement. Verapamil and propranolol, which slow the atrial rate, have no effect.

An interesting discovery of this work is the contradiction of the long-held view that ectopic beats with a left bundle branch block pattern always originate in the right ventricle. At least 50% of these apparently come from the left ventricle. It is still true that ectopic beats with a right bundle branch block pattern coming from the left ventricle.

Dr. Paul Taboul (Univ. of Lyon, France) spoke on the medical management of arrhythmias as affected by some newly available agents. Disopyramide is a new drug having class I effects—it decreases conduction in the his-purkinje systems and in the accessory pathways. Its advantages over other class I drugs are much decreased incidence of side-effects compared with quinidine and a more prolonged action than procainamide (which must be taken every three hours to be effective).

A new class of anti-arrhythmic drugs is the "calcium blockers" i.e., those that interfere with calcium's contribution to the action

potential. The only drugs in this class so far are verapamil and amiodarone, neither of which is currently available in the US. These decrease A-V nodal conduction and increase the A-H interval. Amiodarone appears to be the best available drug for Wolff-Parkinson-White syndrome.

Beta-blockers are another available class of agents that decrease automaticity and A-V nodal conduction and increase action potential duration and refractory periods. Digitalis is also available for supraventricular arrhythmias by decreasing A-V nodal conduction.

In spite of the panoply of drugs now available and the new electrophysiologic knowledge of arrhythmias, a rational approach has not yet been developed for all arrhythmias, and treatment remains empirical—trying drugs on a case by case basis.

Surgical treatment of arrhythmias in cases refractory to medical treatment has been done at a few research centers for about a decade. Now with the dissemination of electrophysiology laboratories, the capability for doing operative treatment has expanded both geographically and in the types of patients who can benefit. Dr. Leonard Horowitz (Univ. of Pennsylvania) detailed these developments.

There are currently four types of arrhythmia which may qualify for this approach—pre-excitation syndromes (the original category), SVT due to concealed bypass tracts, SVT rapid A-V nodal conduction, and most recently ventricular tachycardia. Pre-operative evaluation must include complete electrophysiological study to determine the exact mechanism of the arrhythmia, and full endocardial mapping to determine its exact location. Then surgery is undertaken only when all available drugs and pacemaker therapy have failed to control the arrhythmia.

Dr. Hein Wellens of the Netherlands spoke on "New Concepts in the Pre-excitation Syndromes." He described a recent search for patients over 60 years old with WPW. Very few were found, suggesting that they either die or lose the accessory pathway. Amazingly enough, the answer as to which occurs is not available.

The rhythms manifested by a series of 157 patients studied were SVT in 108, A-fib in 31, and both SVT and A-fib in 10. Electrophysiologic studies have shown that an episode of SVT will start by an ectopic impulse taking advantage of the A-V node's refractory period being shorter than that of the accessory pathway. The impulse thus shoots down the A-V Node and then up the accessory pathway. Some episodes of anti-dromic conduction, whereby the impulse goes down the accessory pathway and up the A-V node, are occasionally seen. These are rare, however.

It has been found that if atrial fibrillation occurs, the shortest R-R interval is very close to the duration of the refractory period of the atrial pathway. It has also become apparent that patients with a short refractory period in the accessory pathway are at high risk if

atrial fibrillation supervenes. Prior to that time, the presence of a short refractory period can be elucidated in the lab by the injection of ajmaline (a new class I anti-arrhythmic not yet available in the US). Failure to achieve a complete block in the accessory pathway by injection of 50-mg ajmaline is highly suggestive of a short refractory period and high risk.

Treatment involves prevention of the arrhythmias: mainly by the use of class I anti-arrhythmics and beta blockers to prevent premature beats and block circus movements. Treatment of an established SVT is the same as for patients without pre-excitation carotid sinus massage, digitalis, beta-blockers, and verapamil.

Cardiomyopathies

Dr. John Goodwin (Royal Postgraduate Medical School, London) spoke on anatomic and physical data recently used to classify the primary cardiomyopathies (diseases of the heart muscle) and their management. These are divided into four types: hypertrophic, congestive, obliterative, and restrictive. Hypertrophic and congestive together comprise about 90% of the total of primary cardiomyopathies in developed countries. Obliterative myopathies are primarily cases of endocardial fibroelastosis, which is found almost exclusively in the tropics. This is characterized by progressive ventricular obliteration and obstruction to inflow. The restrictive myopathies overlap in pathological spectrum with the obliterative and behave similarly in clinical aspects as well.

As the interest of American and European cardiologists is on the first two types, these were the only ones discussed. Primary congestive cardiomyopathies have been an etiologic puzzle. Recent research has revealed a strikingly high proportion—50%—with high titers of antibody against coxsackie virus. This has raised the speculation that antiviral chemotherapy would be useful for this problem. Present therapy is that of conventional medicine for heart failure, with anticoagulants to prevent thrombotic manifestations. Prolonged bed rest, of the order of three to six months, is controversial. It still has its proponents, but the known metabolic derangements associated with it are adequate argument to many that it should not be recommended.

Experiments are being done with inotropic agents, such as glucagon and salbutamol, results of which are still mixed. Vasodilator therapy has been useful to many patients, whether with phentolamine, nitrates, or hydralazine. The often quoted feared side-effect of hydralazine, tachycardia, has not been seen in congestive cardiomyopathy patients. If patients have arrhythmias, however, they may get worse on hydralazine.

Hypertrophic cardiomyopathy has been a subject of intense interest to cardiologists for at least a decade, perhaps out of proportion to its overall incidence in the spectrum of heart disease. Most cases of obstructive myopathy fall into the category called IHSS (idiopathic hyper-

trophic subaortic stenosis). Ultrastructural appearance in this disease shows that the muscle myofibrils are not parallel, but are rather arranged at right angles to one another. It has not yet been determined whether this is completely specific for this disease; but it has prompted some investigators to insist that heart muscle biopsy is necessary to diagnose the disease definitively.

Symptoms and signs in IHSS do not correlate well with the degree of outflow obstruction. The term asymmetric septal hypertrophy (ASH) has been coined to describe patients who have evidence of the morphologic abnormality but no significant obstruction. Clinically these patients may have an S_4 sound, and a systolic murmur which doesn't change on provocative maneuvers. There is a rapid carotid upstroke and a double apical systolic impulse. If obstruction is present, the systolic murmur will alter on provocative maneuvers and there are often both S_3 and S_4 sounds. There is frequently, however, a diastolic murmur as well. The rapid carotid upstroke and double apex beat are present just as in the nonobstructive case.

Provocative maneuvers to evaluate the murmur were outlined. Measures to decrease preload (Valsalva maneuver, standing, exercise, and nitrates) will increase the murmur. Anything that increases contractility (exercise, post-extrasystolic potentiation) will also increase the murmur. Decreases in afterload (vasodilators, nitrates) will also increase the murmur. Anything that increases preload (e.g., squatting) will decrease the murmur.

The clinical course of patients with IHSS is marked by a high frequency of sudden death. This has been attributed to the hemodynamic obstruction. Recent work using Holter monitoring suggests a high frequency of arrhythmias, especially atrial fibrillation and ventricular ectopic beats. These may be far more important as a cause of sudden death than has been realized.

Dr. Alan Pearlman (Univ. of Pennsylvania) reported his series of patients who experienced sudden death. His group has estimated that this occurs at a rate of 5% annually in the population of patients with IHSS. He has collected a series of 25 patients so far, 19 males and 6 females, whose average age of death was 18 years. The typical event occurred after heavy exertion, and came completely without warning. The incidence of this happening does not correlate with the degree of outflow tract obstruction nor with left ventricular end diastolic pressure. In the 18 of the patients on whom EKGs were done prior to death, all had EKG changes. Left ventricular hypertrophy was present in 15, and Q waves in 9.

It has also been noted recently that the cases of sudden death tend to occur in families. Pearlman terms these "malignant families," and arbitrarily defines them as families with two or more first-degree relatives experiencing sudden death. The patients are usually under 25 and are usually asymptomatic or only mildly symptomatic. They seem to be at high risk with severe septal thickening or marked EKG changes.

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Pearlman recommends the following treatment for patients with obstruction: Give propranolol for symptoms—whether pain, syncope, or PVCs; diuretics should only be used for congestive heart failure; if atrial fibrillation supervenes, anti-coagulants should be given; SBE prophylaxis is recommended; surgical myomectomy may be indicated for relief of obstruction of the outflow tract if propranolol is not successful; for hypotensive emergencies, Phenylephrine should be used. Surgical therapy should also be considered in patients in malignant families and in those who survive collapse that would have been fatal without resuscitation.

