Manual of Clinical Problems in Internal Medicine

Annotated with Key References
Second Edition

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Foreword

William Osler was appointed physician-in-chief to The Johns Hopkins Hospital in 1888. One important element of success in the new clinic that he organized was the arrangement for a graded staff, particularly for a graded, full-time, resident staff, among whose members the responsibilities of the day-to-day work with patients were divided. The resident physician, the assistant resident physicians, and the medical interns lived in the hospital and were always in close contact with the work by day and as far as was necessary by night. The resident staff of the clinic consisted of two groups: a lower resident staff of medical interns appointed for a single year and an upper resident staff made up of the resident physician and several assistant resident physicians. These usually were men of exceptional promise; men who had already served as hospital interns and who were willing to enter into a more or less prolonged resident service, often for several years, in order to secure the best possible training for the "higher walks of internal medicine." The position of chief resident physician, which carried with it large responsibilities and opportunities, was a prize to be won only by men of exceptional ability, extensive experience, and favorable promise. The careers of these men during their terms of service subsequently illustrated, on the one hand, the wisdom of their selection and, on the other, the growth-promoting influence of the duties and authority attached to the office.

These words above are paraphrased from a description of Osler as chief of the medical clinic written by Lewellys F. Barker and published in July 1919 in the *Johns Hopkins Hospital Bulletin*. Over the years the residency system described has not only grown at Johns Hopkins but has also furnished the basic pattern for postgraduate training in all medical school hospitals. Its basic premise is that at all times the students and the members of the house staff in the early phases of their training have someone available with more experience than they have to act as sounding boards for their diagnostic analysis of the cases for which they are responsible.

Over the years at Johns Hopkins, the residents have also played a most important part in the training of the medical students, who act as clinical clerks on the inpatient service. This practice has been a valuable resource from the point of view of the students and a very worthwhile experience in learning the art of teaching for the residents.

Formal clinical rounds by the two chief residents are a basic part of the clinical teaching program on the medical service. In this book, the authors' discussions of pertinent clinical problems accompanied by annotated references demonstrate very effectively the usefulness of these rounds in the teaching program. They also provide a concise distillate of the mass of information transmitted in the setting of clinical problems as they are admitted to the clinic. The wealth of clinical material available is well illustrated by the complete nature of the cases presented.

Historians analyzing the various pioneer achievements in Osler's clinic must certainly give high marks to the important influence on American medicine of the residency training program that he developed. This volume represents a worthwhile derivative of one function of the chief medical resident.

A. McGehee Harvey, M.D.

Preface to the Second Edition

The response to the first edition of this volume was very gratifying to us. We are, however, less pleased in retrospect about the quality of our prose. In this second edition, we have endeavored to correct that deficiency. The text and bibliography have been substantially revised, both from the point of view of style and clarity and also in response to advances in medical knowledge over the past three years. We have also abandoned our initial conservatism with respect to the topics selected, a conservatism dictated solely by uncertainty over the size of the original manual, and have added 14 new topics, bringing the total to 165. In addition, we have expanded the annotated bibliography, which now contains more than 3800 references.

Once again we welcome comments and criticisms from our readers. They have been both educational and extremely helpful to us in preparing the second edition.

J. L. S.
H. V. B.

Preface to the First Edition

Resident rounds are the core of the teaching program on the medical service at The Johns Hopkins Hospital. Held three times each week, these rounds are used to review the pathophysiology, differential diagnosis, and management of medical illnesses, with current cases serving as illustrative examples. In order to provide the house staff with background material for these sessions, it has been customary to provide both a discussion and annotated references for each topic reviewed. This practice has been popular with both house staff and students, and it seemed likely that physicians elsewhere who are initiating their training in internal medicine would find this material worthwhile.

Accordingly, we have provided in this volume a concise discussion with annotated references for each of more than 150 clinical problems selected from our individual rounds. We have generally avoided topics of a broad nature and those that are dealt with extensively in medical school curricula. Major emphasis has been placed on pathophysiology and differential diagnosis. Details of therapy have not been stressed since these are well described elsewhere. We have tried to limit our references to journals readily available to the house officer, but have not hesitated to cite more obscure references when they were of merit. When several articles were available on the same subject, we have usually selected the one that seemed most comprehensive or had the best bibliography. We hope that all our omissions are intentional and we welcome any comments or criticisms on articles or topics selected or omitted.

The beginning physician is faced with the problem of absorbing medical tradition as well as with keeping up with the current literature. In the latter instance he can peruse his journals as he receives them. To assimilate the vast quantity of material that has already been bound and shelved is a much more difficult task. With this volume we hope to alleviate that difficulty.

We are profoundly grateful to our mentor, Dr. A. McGehee Harvey, for the opportunity to serve as chief residents in medicine, and to Dr. Philip A. Tumulty for advice and encouragement during the preparation of this volume. The congenial secretarial assistance of Mrs. Nancy Dietz is acknowledged with appreciation.

J. L. S.
H. V. B.

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Cardiac Arrest and Resuscitation

Cardiac arrest may be defined as the sudden, unexpected cessation of effective cardiac output, which, unless reversed, results in death. Sudden, unexpected death occurring outside the hospital is most common in persons with coronary artery disease. Although most of these persons have symptomatic disease, few have new or changing chest pain prior to the arrest; nonspecific symptoms, such as increasing fatigue, irritability, and dyspnea, take on significance only in retrospect. Even when a significant change in symptoms occurs, it is often too closely associated with the cardiac arrest to be of benefit. Sudden death usually occurs at home; ventricular fibrillation is the most common arrhythmia and in those who are resuscitated, myocardial ischemia, particularly of the anterior wall, is as common as acute myocardial infarction. Patients at risk from sudden unexpected death or cardiac arrest in the hospital include those with cardiac disease or respiratory insufficiency, particularly those with the pickwickian syndrome, those with pulmonary emboli, electrolyte or acid-base imbalance, or uremia and those undergoing anesthesia or cardiac catheterization. Patients requiring mechanical ventilatory assistance are at especially high risk. All too often we have observed patients attached to mechanical respirators, with neither the patient nor the respirator ventilating. Ventilator tubing is easily obstructed, and not infrequently patients are intentionally disconnected from the ventilator for repositioning, a practice that cannot be condoned. Failure to flush the lungs with oxygen before tracheal suction may result in a fatal arrhythmia. Patients attached to monitors or ECG machines or with transvenous pacemakers are at risk from ventricular fibrillation due to electrical current leaks. Sudden death can be precipitated by administration of drugs, particularly hypotensive agents or contrast material. Recently, aminophylline was found to be responsible for a large percentage of cardiac arrests in patients without severe underlying disease. This agent is also often responsible for chaotic atrial rhythms as well as ventricular arrhythmias.

A sharp blow to the precordium is the first therapeutic maneuver in resuscitation and the patient's response or lack of one confirms the diagnosis. The precordial blow may abort ventricular tachycardia or convert the asystole of a Stokes-Adams attack. It does not, however, affect ventricular fibrillation.

Attention is then directed to obtaining an adequate airway and promoting circulation by external cardiac massage. The patient must be on a firm surface. Early endotracheal intubation may mean the difference between success and failure, but valuable time should not be wasted in intubation by the unskilled. Mouth-to-mouth or mouth-to-nose ventilation, with occlusion of the other orifice, is an effective temporizing measure. In this regard, insertion of a large-bore needle into the trachea for purposes of ventilation is a useless measure. It should also be recognized that there is no need for ventilation and chest massage to be synchronized. It is more important for the ventilator to keep the patient's neck extended and mandible elevated in order to maintain a patent airway. Only after cardiac massage and ventilation have been established should the type of cardiac rhythm be determined and specific therapy instituted. *It cannot be emphasized too strongly that the simple techniques just described provide the necessary stabilization required for more specific therapy to be effective and that if they are not performed properly, a successful outcome is unlikely.* The femoral pulse rate and the pupillary size, if not compromised by drug therapy, can be used to judge the adequacy of the resuscitative effort. Inadequate circulation is frequently due to failure of the person performing the massage to position his weight

directly over the sternum or to an early release of sternal compression. Improper positioning results in complications, such as rib fractures and ruptured viscera. If the chest is not compliant, particularly in patients with intracardiac pacemakers or after cardiac catheterization, the possibility of pericardial tamponade should be considered.

As soon as possible, oxygen should be administered, and intravenous sodium bicarbonate if necessary, to combat acidosis and hyperkalemia, which depress cardiac contractility and increase cardiac irritability. Excessive bicarbonate administration must, however, be avoided because it promotes an increase in serum osmolality and arterial P_{CO_2}. Gastric dilatation, which may impair ventilation and contribute to aspiration and mucosal laceration, can be avoided by passage of a nasogastric tube. Unless the terminal cardiac rhythm is known, immediate electrical defibrillation is not indicated, since asystole is not uncommon and restoring circulation by cardiac massage is more important. Once that is achieved, specific therapy can be directed by electrocardiographic findings. There is often reluctance to administer epinephrine by intracardiac injection during resuscitation, but in an "all or none" situation such conservatism is inappropriate. If properly performed, the procedure can provide a potent mechanical stimulus to cardiac contraction as well as assure access of the drug to the circulatory system.

The outcome of cardiac resuscitation depends on many factors, including the nature of the underlying disease, the cause of the arrest, and the duration of hypoxemia before initiation of therapy. Resuscitative efforts are less likely to be successful in patients with congestive heart failure, respiratory failure, shock, or coma. Postresuscitation bradycardia and junctional or idioventricular rhythms are also associated with a poor prognosis. When ventricular fibrillation unassociated with myocardial infarction is the cause of cardiac arrest, there is a high incidence of recurrent fibrillation, often without warning, in the survivors. Many of the persons so affected have been found to have surgically remediable coronary artery disease.

The duration of resuscitative efforts is a matter of individual decision. If a spontaneous pulse or an adequate electrically stimulated pulse is not achieved within two hours, a successful outcome is unlikely. Often electrocardiographic activity returns without mechanical activity. The electromechanical dissociation may be due to cardiac rupture, pump failure, tamponade, or pulmonary embolism.

Following successful return of cardiac activity, the patient must be evaluated for the cause of the arrest as well as for complications resulting from it (among them, pneumothorax, aspiration, rib fractures, marrow emboli, visceral injury, neurologic deficits, cerebral edema, and renal tubular necrosis). (J.L.S.)

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Dissecting Aneurysm of the Aorta

Dissecting aneurysm is the most common acute disease of the aorta. Dissection can occur at any age but most develop between the fifth and seventh decades, with a predominance in men. Factors predisposing to aortic dissection include atherosclerosis, hypertension, cystic medial necrosis, giant cell arteritis, Marfan's syndrome (but not homocystinuria), idiopathic kyphoscoliosis, coarctation of the aorta, Turner's syndrome, aortic hypoplasia, bicuspid aortic valves, and pregnancy. To date, dissection has not been reported in patients taking oral contraceptives. Syphilis does not protect against dissection, nor does hypothyroidism predispose to it.

Dissections can be divided into three types based on the location of the intimal tear. Type 1 dissections, the most common, begin in the ascending aorta and extend proximally and distally to involve the whole aorta, usually in an asymmetric fashion following the outer aorta wall. Type 2 dissections begin in the ascending aorta or arch, are limited to that area, and are most often associated with Marfan's syndrome. Type 3 dissections begin beyond the left subclavian artery and extend distally. Involvement of aortic branches is not uncommon, but it is usually limited to those arteries with more elastic tissue than muscular tissue. They include the iliac, innominate, and subclavian arteries.

The factors responsible for aortic dissection are not totally understood. Defective connective tissue is likely in Marfan's syndrome, coarctation, bicuspid valves, cystic medial necrosis, aortic hypoplasia, idiopathic kyphoscoliosis, and, possibly, pregnancy. On the other hand, many normal aortas show medial degeneration, and it is not clear whether the medial degeneration found in patients with dissections is the cause of the process or

a result of it. Because intimal tears are not always found, dissection has been thought to result from an intramural hematoma in these patients. It is unlikely, however, that the vasa vasorum could generate the necessary pressure relative to the aorta to produce such a hematoma. Hypertension is also not an invariable feature, and the aorta can withstand intramural pressures well above those associated with dissection. Atherosclerosis may play a more significant role than is generally accepted. The process produces damage to the media as well as the intima, and a ruptured plaque can provide the necessary channel for the dissection. In addition, atherosclerosis-induced rigidity and dilatation of the aorta can result in pressure changes (Laplace's law) that may favor dissection. The importance of rigidity is reflected by the area of location of intimal tears, all of which are at points of fixation of the aorta.

Propagation of the dissection seems to be aided by the pulsatile nature of aortic blood flow, suppression of which is the major objective in the medical management of the illness. Propagation is also aided by the anatomy of the aorta, with its lamellation and vasa vasorum. Most commonly, the dissection occurs in the area between the outer third of the media and the adventitia. As a consequence, there is little support for the false channel, and the result is often rupture into the pericardium, mediastinum, or pleural space. Spontaneous reentry can occur, but it does not protect against rupture of the false lumen.

The onset of aortic dissection is usually abrupt, with the development of pain in the chest or abdomen. Although pain limited to the back is distinctive for type 3 dissection, there is otherwise no relationship between the location of the pain and the intimal tear, nor is there any feature of pain that might distinguish a dissection from the other diseases that must be considered in a differential diagnosis. Those diseases include acute myocardial infarction, pancreatitis, perforated peptic ulcer, leaking aortic aneurysm, vertebral collapse, esophageal rupture, and acute cholecystitis. Because of the nonspecific nature of its presentation, aortic dissection must be considered in every acute thoracic and abdominal syndrome. Factors suggesting dissection are chest or abdominal pain associated with loss of one or more peripheral pulses, pericardial tamponade, hemothorax, and neurologic signs. The neurologic signs include sudden coma or syncope, hemiplegia, extraocular paralyses, Horner's syndrome, vocal cord paralysis, neuropathy, and seizures. Either the right or both cerebral hemispheres can be involved, but not the left one alone. The neurologic signs are the result of either hypotension (pericardial tamponade) or arterial occlusion by the dissection. Differences in blood pressure between the extremities or loss of a pulse are important physical signs, as is the development of aortic insufficiency. An uncommon but helpful sign is a prominent pulsation of either sternoclavicular joint. The ECG is helpful only to exclude an acute myocardial infarction but abnormalities suggesting myocardial injury are seen in 20 percent of dissections. A plain chest x-ray is never diagnostic since aortic widening due to dissection cannot be distinguished from mediastinal fat, hematoma, tumor, or aortitis; aortography is necessary for the diagnosis and classification of the type of dissection. The procedure is not without risk, but the hazards of missing the diagnosis outweigh the risk.

Studies of the natural history of aortic dissection have indicated that 3 percent of patients die immediately, 21 percent within 1 day, 60 percent in 2 weeks, and 90 percent in 3 months. Factors predisposing to a high mortality include the presence of underlying disease, severe neurologic involvement, and type 1 or 2 dissection. Type 3 dissections and failure to opacify the false lumen at aortography are associated with a good prognosis.