PATHOLOGY of the HEART

Edited by

S. E. GOULD, M.D., D.Sc.

Clinical Professor of Pathology, Wayne University College of Medicine Detroit, Michigan

> Pathologist, Wayne County General Hospital Eloise, Michigan

Consultant in Pathology, Veterans Administration Hospital Dearborn, Michigan

Editor, American Journal of Clinical Pathology



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Collaborators

ARCHIE H. BAGGENSTOSS
Associate Professor of Pathologic Anatomy
Graduate School, University of Minnesota
Section on Pathologic Anatomy, Mayo Clinic
Rochester, Minnesota

ALEXANDER BARRY
Associate Professor of Anatomy
University of Michigan Medical School
Ann Arbor, Michigan

JESSE E. EDWARDS
Assistant Professor of Pathologic Anatomy
Graduate School, University of Minnesota
Section on Pathologic Anatomy, Mayo Clinic
Rochester, Minnesota

SYLVESTER E. GOULD
Clinical Professor of Pathology
Wayne University College of Medicine
Detroit, Michigan
Pathologist, Wayne County General Hospital
Eloise, Michigan

HAROLD D. GREEN
Professor of Physiology-and Pharmacology
Bowman Gray School of Medicine
Winston-Salem, North Carolina

WILLARD T. HILL Associate Professor of Pathology Northwestern University Medical School Chicago, Illinois

EDWARD B. KRUMBHAAR
Professor of Pathology (Emeritus)
University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania

ALAN R. MORITZ
Professor of Pathology
Western Reserve University School of Medicine
Cleveland, Ohio

GORDON B. MYERS
Professor of Medicine
Wayne University College of Medicine
Detroit, Michigan

BRADLEY M. PATTEN
Professor of Anatomy
University of Michigan Medical School
Ann Arbor, Michigan

Otto Saphir

Clinical Professor of Pathology University of Illinois College of Medicine Pathologist, Michael Reese Hospital Chicago, Illinois

Walter A. Stryker
Assistant Professor of Pathology
Wayne University College of Medicine
Detroit, Michigan
Pathologist, Wyandotte General Hospital
Wyandotte, Michigan

WILLIAM B. WARTMAN
Professor of Pathology
Northwestern University Medical School
Chicago, Illinois

To My Wife

Foreword

M OST PATHOLOGISTS look on their subject as a large segment of the whole field of medicine, whereas some regard it as an independent science for the investigation of basic biological phenomena. Naturally, these viewpoints are not mutually exclusive, a fact that is well exemplified in this volume. The practical aspects of the subject predominate, so that clinicians and pathologists will find a comprehensive survey of the information now available.

The book demonstrates beyond question that pathology in America has grown up; it has reached a stage of maturity which gratifies all its devotees. Any scientific presentation is benefited by an appreciation of its historical background, especially when, as in this instance, the various aspects of pathological anatomy, disordered function, and clinical manifestations are well covered. The same approach is manifest in all the discussions, whether of embryology, normal anatomy, etiology, or any of the diseased states. Thus is integrated in each chapter that broad concept of cause, form, and function which modern pathology requires. The material is carried over into the clinic by clear and adequate discussions of diagnosis and rational treatment. Moreover, the deficiencies of our present knowledge are so portrayed as to open many fields for future research.

The editor is to be congratulated on his choice of contributors and the selection of subjects. The authors of the various sections can be felicitated on the orderly, scientific, and thorough coverage of the fields in which they

are unquestioned experts.

This book is a superb contribution to the literature of pathology, medicine, and biology, and should be welcomed in the special field of cardiology. It is to be anticipated that its reception will warrant repeated editions with opportunities for such revisions as the advances of science justify.

HOWARD T. KARSNER, M.D.

Preface

TP TO THE PRESENT time there has been no adequate book embracing the pathology of the heart in the English language. In undertaking the preparation of such a volume the authors and the editor have attempted to present the essential factual data in a practical and understandable manner, not only for the pathologist but also for the clinician, and perhaps also for the medical student.

A concise introductory chapter presents a restrained historical review of the subject in perspective. Next follow three chapters embodying the pertinent present knowledge of the embryology, the anatomy and the normal and pathologic physiology of the heart. The body of the treatise is devoted to the available authentic knowledge of cardiac pathology. And finally, a chapter on clinicopathologic correlations supplements remarks on such correlations made within the content of the separate chapters on pathology. The emphasis throughout on the clinical aspects of cardiac pathology is deliberate since, in the last analysis, the lessons of pathology must be brought into relation with the living patient.

In order to avoid overlapping of discussion of the same topics by different authors it will be found that, although particular subjects may be mentioned at several points, in practically all such instances the subject has been given a relatively full discussion at one point only.

In preparation of this work, the editor has entertained the hope of realizing two objectives: first, that the data presented, so far as is possible, be accurate and authentic, and second, that all collaborators upon completion of their work regard their participation in this joint endeavor as having been worthy of the effort. The reader, however, must be the final judge of the worth of this volume. In this connection the editor will welcome any advice, criticism or suggestion for improvement of this monograph, should the reception of this first edition warrant the publication of a second edition.

It is a pleasure to acknowledge the splendid cooperation which the editor has received both from his collaborating authors and from the publisher.

Grateful acknowledgment is made to Dr. Carl V. Weller, Professor of Pathology, University of Michigan, for assistance rendered in planning the organization of this book, and to General Raymond O. Dart, former Director, and Dr. Webb Haymaker of the Armed Forces Institute of Pathology for the use of illustrative material. Sincere appreciation is expressed to the many who in confidence have critically reviewed manuscripts of the several chapters; to Mrs. Roslyn K. Stempel, my former secretary, for conscientious review of manuscripts and preparation of copy, and to Miss Edith E. Parris, Assistant Editor of the American Journal of Clinical Pathology, for advice and assistance in review of page proof. The editor also wishes to acknowledge his obligation to Dr. Roland M. Athay, Superintendent of Wayne County General Hospital and Infirmary, and to the

Wayne County Board of County Institutions, the governing body of the hospital, for their public-spirited support of this undertaking. And finally he wishes to offer a word of tribute in remembrance of Dr. Thomas K. Gruber, late Superintendent of this hospital, a wise and able administrator, and a true and beloved friend who gave encouragement to this task, as indeed he did to every worthy endeavor.

S. E. GOULD

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PATHOLOGY of the HEART

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History of the Pathology of the Heart

EDWARD B. KRUMBHAAR

Cardiac Rupture	Congenital Heart Disease	
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Useful knowledge of the pathology of the heart - by "pathology" being meant all the changes produced by disease - is a relatively recent acquisition along the long path of medical history. The slowly accumulating knowledge of disordered cardiac structure and function produced little of practical value before the nineteenth century. Ancients even maintained that the heart was not subject to disease - Cor non aegrotare posse, as Hippocrates is said to have put it.* Galen's classification of types of heart disease (wounds and inflammations, pericarditis and pericardial effusions, palpitations) held the field for more than a thousand years. It was only in the fifteenth century that some of the grosser anatomic changes began to be observed. Pietro di Montagnano (died 1460), for instance, noted damaged hearts in 14 dissections at Padua.

Functional disturbances such as "palpitation" (frequently considered by Hippocrates) and arrhythmia (a term attributed to Galen) were naturally recognized much earlier, though interpretation of the former word at least was very different then from what it is now, and both were understood so vaguely that they contributed nothing of true value. In fact, an irregular pulse amounted to little more than an irregular pulse until instruments of precision * led to classification of the arrhythmias into recognizable types of greatly varying causation and significance (J. Mackenzie and Thomas Lewis). While consideration of these functional advances. which are of prime importance in the development of the recently established specialty of cardiology, may be left to the physiologist and clinician and to histories of the general subject, pathologists should remember and teach that they also are truly developments in the field of pathology, even if they were not chiefly contributed by professional pathologists. The same principles apply to other less obvious examples of cardiac pathologic physiology, such as palpitation in the modern sense, "irritable heart" (J. M. da Costa, 1867);

^{*} This statement is offered by both Moon and Herrick as the basis for the notion that the heart cannot be diseased. The nearest that I, with the help of W. B. McDaniel II, have come to this notion is in Littré's translation of de Morbis: "Nullus in corde morbus suboritur" (No disease arises in the heart); elsewhere it is stated that the heart does not labor with pain. It is suggested that "aegrotare" may be an early paraphrase of the original Greek, in which case Hippocrates should not be held responsible for the more glaring error.

^{*} Development of graphic instruments of precision in this field is so closely linked with growth of knowledge of the pathologic physiology of the heart beat that one should notice especially such advances as Vierordt's (1854), Marey's (1860), and Dudgeon's (1882) sphygmographs; Potain's (1867), Mackenzie's (1902) polygraphs; Frank's (1907) capsule; Waller's cardiac electrometer (1887); and Einthoven's electrocardiograph (1903).

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Tabula xx11. ad Observationem Lx1x. cor triplici ventriculo præditum ostendit.

- A. Cor triplici præditum ventriculo.
- B. B. Duo dextri cordis pentriculi.
- C. Sinister ventriculus.
- D. Arteria pulmonaria ex utroque dextro ventriculo prodiens.

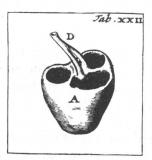


Figure I-1. Kerckring's report (1670) of a heart with double right ventricle and pulmonary artery, showing the crudity of most 17th century illustrations. N.B. The letters BB and C do not appear in the original illustration.

cardiac pain (Heberden, 1768); cardiac murmurs, gallop rhythm (Bouillaud, 1847; Potain, 1887); cardiac dyspnea (Cheyne, 1818; Stokes, 1846); and cardiac edema.

Knowledge of the pathologic anatomy the basis for comprehension of almost all pathologic phenomena - of the heart, grew slowly. Its development conveniently falls into three overlapping periods: the first, longest and least valuable, characterized by isolated observations of such material as chance offered; next, a 300-year period of systematized collections of clinicopathologic cases; and third, in the nineteenth and twentieth centuries, treatises and textbooks on the ever-increasing body of known facts and their underlying laws. Negligible through ancient, classical, and medieval times, the practical beginning of cardiac pathology may conveniently be located in the Renaissance with Benivieni's De abditis causis morborum (published

posthumously, 1507). Of his 111 short chapters, one of the 20 with necropsy describes what seems to be an acute pericarditis (it is not clear whether by the cavity "cum pilis refertum" - stuffed with hairs - is meant that of the pericardium or of the cardiac chambers). Another case revealed a "polyp of the heart," a subject which remained in confusion well into the nineteenth century. However, the heart was long a minor contributor to the progress of the anatomic concept of disease. Even in the great Sepulchretum (1679) of Bonetus, a systematic presentation of nearly 3000 clinicopathologic cases from the literature, we find few significant items of cardiac pathology, though there are dozens of cases of "palpitation" and a number of the so-called cardiac polyps, some even being thus diagnosed before death. Bauhin (1592), Tulp (1641), and Malpighi (De polypo cordis, 1686) were others who prolonged this fallacy, until Kerckring showed the dark red, easily removable kind of polyp to be merely a postmortem phenomenon (Spicilegium anatomicum, 1670, Obs. 73). The error was only gradually eliminated as the gray, tightly adherent antemortem thrombus became differentiated from the loose red postmortem clot. Even in 1839, in the first edition of Gross' excellent Elements, polyps appeared as "polypous concretions," though in the third edition (1857) "polypous" was changed to "fibrinous." Kerckring also contributed to the pathology of congenital heart disease; his 69th observation showed an infant's heart with a double right ventricle (cor triplici ventriculo) and a pulmonary artery leading from each, to fuse later, as is well shown in his engraving (Figure I-1).

In the seventeenth century, with the increasing accumulation of single pathologic observations, the custom arose of publishing assembled cases, whether from the

literature or from personal observation, collections that at times reached large numbers. Thus, Bonetus in his Sepulchretum included 2934 observations (i.e., cases) made by 470 authors, yet he reports very little about the heart. One section, De angina, includes only five observations, and these are mostly not pectoris. These collections were conspicuously weak in their heart material, and even Morgagni's great De Sedibus et Causis Morborum, which is credited with generally establishing the anatomic concept of disease, contained much less about the heart than about other organs and systems. However, we find reported the structural changes in a case of angina pectoris, one of the earliest accounts of heart block, cases of vegetative endocarditis - one associated with gonorrhea (other names, of course, being applied), a case of rupture of the heart, a case of congenital hypoplasia of the aorta. Morgagni had the great



Figure I-2. Giovanni Battista Morgagni (1682-1771), by Angelica Kauffmann. (From Castiglioni's Storia di Medicina)

merit of correlating careful clinical study with autopsy findings, better than any of his predecessors and, for some years, of his successors as well. One of the greatest medical figures of the eighteenth century, he (Figure I-2) also made a noteworthy contribution to knowledge of the pathology of the heart.

Systematic descriptions of cardiac pathology, though not comparable to modern textbooks, had already begun to appear. An early example was Fernel's Pathologia, one of the chief divisions of his great Universa Medicina (1554). In it he grouped heart diseases under the peculiar headings of inflammation, erysipelas, tumores contra naturam, ulcer and wounds. Vieussens' Traité Nouveau de la Structure et des Causes du Mouvement du Coeur (1715), on the other hand, was "the first to make serious contributions to our knowledge of diseases of the heart" (Moon). He dealt with true lesions (congenital anomalies, mitral stenosis and aortic regurgitation, pericardial inflammation, adhesions and effusions) and not with symptoms that were but vaguely comprehended. He recognized that asthma and hydrothorax might be due to heart disease, and also had a good concept of the passive congestion resulting from valvular obstruction. Lancisi (1728), who, according to Long, "laid the foundation for a true understanding of the heart," observed sclerotic and warty valves, and dwelt at some length on cardiac "aneurysms" (in the strict etymologic derivation of dilatation, a meaning that still applies to some varieties). Thus to him aneurysms were commoner in the atria than in the ventricles, and were least common in the left ventricle. His handling of the etiology of heart disease was less successful, stressing congenital defects, long-continuing violent emotions or physical effort and palpitations. His contemporary, Albertini, favored

heredity, and syphilis (using mercury in its treatment), as important causes of heart disease, and correlated dyspnea and pulmonary edema with cardiac disease (1726).

Senac's De la Structure, de l'Action et des Maladies du Coeur (1749), however, was the first extensive work devoted to the heart alone. The 11 chapters on Dis-

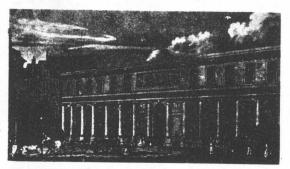


Figure I-3. École de Médecine de Paris in the early 19th century, when French pathology was dominant. (From a contemporary engraving in the Bibliothèque Nationale.)

eases of the Heart, contained much pathology. He recognized inflammations of all three layers, and that pericarditis might follow pneumonia or pleurisy or infectious fevers, that aneurysm (i.e., dilatation) of the ventricle might be accompanied by hypertrophy. He considered tumors, abscesses, "ulcers" and wounds, as well as arrhythmia, palpitations and weak action (syncope).

Textbooks of pathology, in the sense in which we use the term "textbook" today, may be said to have started modestly with Matthew Baillie's small The Morbid Anatomy of Some of the Most Important Parts of the Human Body (1793). The 24 pages of the chapters on The Heart and Pericardium touch briefly on inflammation, abscess, gangrene, polyp, aneurysm of the heart, fibrous, bony, and earthy thickening of the valves, rupture of valves and of myocardium, malformations and hypertrophy of the heart; and "white spot," inflammation, adhesions, dropsy, excessive dryness

and "scrofulous tumors" of the pericardium. No case reports were included, except in the German translation. The early textbooks in France and Germany, such as Lobstein's Traité d'Anatomie Pathologique (1829, 1833) and Meckel's Handbuch (1812-18) dealt with general pathologic anatomy only, the specialized pathology often being covered by atlases. The first textbook of any size in this country, Samuel Gross' Elements of Pathological Anatomy (1839), on the other hand, was divided, as are most texts today, into General and Specialized Pathological Anatomy. In the latter portion, the heart and its membranes occupied only 40 of the 510 pages. Rupture was regarded by Gross, and for a long time afterward, as "generally the result of ulceration or of the softening of fatty degeneration."

The French school, dominant in the early nineteenth century through Europe's political situation and French pioneer achievements in physical diagnosis, also set the pattern in the pathology of the heart, through two great clinicians, Corvi-



Figure I-4. Réné-Théophile-Hyacinthe Laennec (1781-1826). (From a miniature on ivory in the Faculty of Medicine of Paris.)

sart and Laennec. Corvisart's Essai sur les Maladies et les Lesions Organiques du Coeur (1806), though clinical lectures with pathologic checks, established our present custom of considering heart disease according to its three layers (with a fourth class for diseases contra naturam and those affecting several tissues). He emphasized the frequency of organic heart disease - second only to pulmonary phthisis, correctly unraveled the tangled problem of polyps, distinguished between fatty infiltration and degeneration of the heart (the latter he said he had never seen), described rupture of chordae tendineae and papillary muscles, and included the conventionally accepted lesions as well as hydatids. Laennec (Figure I-4) in his Traité de l'Auscultation Médiate (1819 and 1826), largely followed Corvisart; however, with extensive correlation of his clinical and autopsy findings, he was able to attain greater precision in his descriptions. In his consideration of different sizes of the heart. he recognized that heart failure was seldom seen except in dilated hearts, with or without hypertrophy. He had learned that communications between the ventricles were not always congenital malformations. He was accurate in describing irregularities and murmurs, yet he made a false interpretation of the origin of the two normal heart sounds, and although he saw postmortem hemolytic staining of the valves he attributed it to inflammation.

Thereafter, the heart occupied increasing space in textbooks of pathology.

To take up achievements in some of the special fields, we turn first to Congenital Malformations.

Congenital Heart Disease offers some good examples of a long random progress of pathologic knowledge in a given subject before integration. A number of its lesions had been observed and reported for centuries before the observations were coordinated and the knowledge adequately



Figure I-5. Carl Rokitansky (1804-1878). (From an original carte de visite photograph, College of Physicians of Philadelphia.)

utilized. Correlation began in the nineteenth century, as in Farre's Malformations of the Human Heart (1814). Peacock's work of the same name (1858) is the first comprehensive treatise that gathered much from the earlier and the prolific contemporary literature. Yet comparison of his largely empirical explanations of the genesis of the congenital lesions with Rokitansky's (Figure I-5) Die Defekte der Scheidewände des Herzens (1875) shows the rapidity of progress in this field (Figure I-6). This can be ascribed to recently acquired embryologic knowledge, as well as to zeal in unearthing new kinds of malformations. This work also achieved the distinction, seldom reached in the biological sciences, of correctly predicting that certain unobserved cardiac anomalies would later be discovered. Among congenital anomalies Rokitansky's Lehrbuch der pathologischen Anatomie (third edition, 1856) included acardia, ectopia, two-