

(Second Edition)

PATHOLOGY *of the* HEART

Edited by

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

*Professor of Pathology, Wayne State University College of Medicine
Professor of Pathology, University of Detroit School of Dentistry
Detroit, Michigan*

*Director of Pathology, Wayne County General Hospital
Eloise, Michigan*

*Research Associate and Lecturer in Pathology,
University of Michigan School of Medicine
Ann Arbor, Michigan*

With a Foreword by
HOWARD T. KARSNER, M.D.



CHARLES C THOMAS • PUBLISHER
BANNERSTONE HOUSE
301-327 East Lawrence Avenue, Springfield, Illinois, U.S.A.

Published simultaneously in the British Commonwealth of Nations by
BLACKWELL SCIENTIFIC PUBLICATIONS, LTD., OXFORD, ENGLAND

Published simultaneously in Canada by
THE RYERSON PRESS, TORONTO

This monograph is protected by copyright. No
part of it may be reproduced in any manner
without written permission from the publisher.

© 1960 by CHARLES C THOMAS • PUBLISHER

Library of Congress Catalog Card No. 58-12154

FIRST EDITION, 1953

SECOND EDITION, 1960

Printed in the United States of America

Collaborators

ARCHIE H. BAGGENSTOSS

*Professor of Pathology
Graduate School, University of Minnesota
Head, Section of Pathologic Anatomy, Mayo Clinic
Rochester, Minnesota*

CHARLES P. BAILEY

*Professor and Head, Department of Thoracic Surgery
Hahnemann Medical College and Hospital
Philadelphia, Pennsylvania*

ALEXANDER BARRY

*Professor of Anatomy
University of Michigan Medical School
Ann Arbor, Michigan*

JESSE E. EDWARDS

*Professor of Pathology
Graduate School, University of Minnesota
Consultant, Section of Pathologic Anatomy, Mayo Clinic
Rochester, Minnesota*

IRA GORE

*Assistant Clinical Professor of Pathology
Harvard Medical School
Chief of Laboratory Service, Veterans Administration Hospital
West Roxbury, Massachusetts*

SYLVESTER E. GOULD

*Professor of Pathology
Wayne State University College of Medicine
Detroit, Michigan
Director of Pathology, Wayne County General Hospital
Eloise, Michigan*

HAROLD D. GREEN

*Professor and Director, Department of Physiology and Pharmacology
Bowman Gray School of Medicine
Winston-Salem, North Carolina*

MIGUEL A. GUERRERO

*Department of Pathology
University of Alabama Medical Center
Birmingham, Alabama*

PATHOLOGY OF THE HEART

WILLARD T. HILL

*Abbott Associate Professor of Pathology
Northwestern University Medical School
Chicago, Illinois*

JOSEPH E. IMBRIGLIA

*Professor of Pathology
Hahnemann Medical College and Hospital
Philadelphia, Pennsylvania*

EDWARD B. KRUMBHAAR

*Professor of Pathology (Emeritus)
University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania*

MAURICE LEV

*Professor of Pathology
Northwestern University Medical School
Director, Congenital Heart Disease Research and Training Center,
Hektoen Institute
Chicago, Illinois*

AVERILL A. LIEBOW

*John Slade Ely Professor of Pathology
Yale University School of Medicine
New Haven, Connecticut*

JOSEPH F. A. McMANUS

*Professor and Chairman
Department of Pathology
University of Alabama Medical Center
Birmingham, Alabama*

ALAN R. MORITZ

*Director, Institute of Pathology
Western Reserve University
Chief, Pathology Service, University Hospitals
Cleveland, Ohio*

GORDON B. MYERS

*Professor of Medicine
Wayne State University College of Medicine
Detroit, Michigan*

BRADLEY M. PATTEN

*Professor of Anatomy
University of Michigan Medical School
Ann Arbor, Michigan*

COLLABORATORS

OTTO SAPHIR

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

WALTER A. STRYKER

*Associate Professor of Pathology
Wayne State University College of Medicine
Detroit, Michigan
Pathologist, Wyandotte General Hospital
Wyandotte, Michigan*

WILLIAM B. WARTMAN

*Morrison Professor of Pathology
Northwestern University Medical School
Chicago, Illinois*

Foreword

THIS NEW EDITION, as was anticipated from the welcome afforded the first edition, gives ample evidence of the high regard in which the book is held by the profession as a whole and that large component devoted to the study of heart disease. The first edition exemplified a broad approach to the subject in that embryology, normal anatomy and histology as well as physiology and etiology were combined with pathologic anatomy, pathologic physiology and clinical manifestations so as to constitute a unified consideration of current knowledge and of research potential. The same breadth of approach prevails in this new edition. Pathology, in its maturity, continues to grow and develop.

It has been necessary to reset the type completely which adds to the exceptional opportunity for revision by those who contributed to the earlier edition. This situation has been exploited in many ways, such as in sentence and paragraph structure, certain changes in classification and terminology, insertion of new illustrations, alteration of paragraph headings and extension of bibliography. It is fortunate indeed that the original authors are available for the revision, and the enthusiasm of their response is exemplary. A certain amount of alteration has been necessary to harmonize the presentation with new chapters. The use of somewhat smaller type is not a conspicuous change and the book can be read as comfortably as ever.

New chapters expand the overall coverage admirably. The wisdom of the choice of topics is complemented by the selection of contributors known to be recognized authorities in their fields. Catholicity is evident throughout but we may be proud that a group of Americans can write a book of this character. And each new chapter continues the fine tradition of the earlier edition. These follow the methods of the preceding edition in discussion of form and function, of clinical manifestations and diagnosis and of prognosis and therapy.

The chapter on conduction system clearly delineates widely accepted views and presents fairly and adequately the few controversial aspects. The pathology of the aorta is now described sequentially and in a manner readily available for reference. A comprehensive presentation of histochemical procedures is a great value to any pathologist and is well adapted to the special study of the cardiovascular system. In the course of becoming a spectacular adjunct to the treatment of acquired and congenital lesions of the heart, surgery has compelled the pathologist to give special and often instructive attention to the various correctible conditions. The methods, usefulness and limitations of surgical attack are admirably described. The influence of heart disease on other organs and systems of the body has been recognized for a long time and has been carefully studied. Only comparatively recently has the interdependence of cardiac and pulmonary function been firmly established. To have this relationship so well covered that all students of disease in the thorax may profit is a fine addition to the book.

An impressive perspective of the whole subject is provided by the historical review, with amplification in individual chapters, and the suggested avenues for further study and research. The editor and contributors are to be congratulated on a task well done.

HOWARD T. KARSNER, M.D., LL.D.

Preface

THE COLLABORATORS of this monograph on the heart deeply appreciate the cordial reception by the medical public of the first edition. The call for a second edition within the space of six years indicates that the volume has filled a need. It is gratifying to have all of the collaborators of the first edition participate in the preparation of the new edition, and to note that they have done so with even greater enthusiasm.

Owing to the extensive revisions and additions that have been made, it has been necessary to reset the type completely. In addition, five new chapters have been added, covering the following subjects: the conduction system, diseases of the aorta, cardiopulmonary disease, surgery of the heart, and histochemical procedures. The emphasis has remained on correlation of the clinical and pathologic findings. In order to make room for new material included in the original chapters and for the additional chapters, and still keep the selected material within the space of one volume, it has been necessary to limit the space allowances sharply and to use smaller sizes of type.

Sincere thanks and appreciation are gratefully expressed to the collaborators for their splendid cooperation; to Mr. Charles C Thomas and his associates for outstanding achievement in bookmaking; to Mrs. Ruby Arquette for meticulous care in typing and proofreading; and to Mrs. Ruth Tobey for assistance in the preparation of the indices and for proofreading.

Finally, the editor offers the hope that this volume may be found useful and that it may reflect credit to the field of pathology by supplying the reader with readily available authentic information with reference to diseases of the heart.

S. E. GOULD

Contents

	<i>Page</i>
Collaborators	v
Foreword	ix
<i>By Howard T. Karsner, M.D., LL.D.</i>	
Preface	xi
I. History of the Pathology of the Heart	3
<i>By Edward B. Krumbhaar</i>	
II. The Development of the Heart	24
<i>By Bradley M. Patten</i>	
III. The Structure of the Adult Heart	93
<i>By Alexander Barry and Bradley M. Patten</i>	
IV. The Conduction System	132
<i>By Maurice Lev</i>	
V. Physiology of the Heart	166
<i>By Harold D. Green</i>	
A. Normal Cardiac Physiology	166
B. Abnormal Cardiac Function	200
VI. Congenital Malformations of the Heart and Great Vessels	
<i>By Jesse E. Edwards</i>	
A. Malformations of the Atrial Septal Complex	260
B. Malformations of the Ventricular Septal Complex	294
C. Malformations Resulting from Abnormalities in Partitioning of Truncus and Conus Arteriosus	354
D. Malformations of the Valves	375
E. Malformations of Endocardium and Pericardium	417
F. Malformations of the Coronary Vessels	425
G. Aneurysms of the Aortic Sinuses (Valsalva)	435
H. Malformations of Aortic Arch System	438
I. Malformations of the Thoracic Veins	481
VII. Degenerative Lesions	497
<i>By William B. Wartman and Willard T. Hill</i>	
VIII. Diseases of the Coronary Arteries	555
<i>By S. E. Gould</i>	
A. Atherosclerosis	555
B. Non-atheromatous Lesions	634
IX. Rheumatic Disease of the Heart	643
<i>By Archie H. Baggenstoss and Otto Saphir</i>	
X. Nonrheumatic Inflammatory Diseases of the Heart	696
<i>By Otto Saphir</i>	
A. Pericarditis	696
B. Endocarditis (Nonrheumatic)	710
C. Myocarditis	779

PATHOLOGY OF THE HEART

	<i>Page</i>
XI. Parasitic Diseases of the Heart..... <i>By Walter A. Stryker</i>	824
XII. Injuries of the Heart and Pericardium by Physical Violence..... <i>By Alan R. Moritz</i>	843
XIII. Neoplasms of the Pericardium and Heart..... <i>By Otto Saphir</i>	859
XIV. Lesions of the Aorta..... <i>By Ira Gore</i>	896
XV. Cardiopulmonary Disease <i>By Averill A. Liebow</i>	940
XVI. Clinicopathologic Correlations <i>By Gordon B. Myers</i>	983
XVII. Surgery of the Heart..... <i>By Charles P. Bailey and Joseph E. Imbriglia</i>	1015
XVIII. Gross Examination of the Heart. Injection of Coronary Arteries. Weights and Measurements of Heart..... <i>By Otto Saphir</i>	1043
XIX. Histochemical Procedures in the Study of Heart Disease..... <i>By J. F. A. McManus and Miguel A. Guerrero</i>	1067
Author Index	1085
Subject Index	1115

PATHOLOGY
of the HEART

History of the Pathology of the Heart*

EDWARD B. KRUMBHAAR

Early Observations	3	Pericardium	16
Congenital Heart Disease	8	Endocardium	16
Size of Heart	12	Myocardium	18
Coronary Heart Disease	13	Functional Disturbances of Heart Action	20
Cardiac Rupture	14	Cardiac Arrhythmias	21
Rheumatic Heart Disease	14	Bibliography	23
Cardiac Syphilis	15		

USEFUL KNOWLEDGE of the pathology of the heart—"pathology" including all the changes produced by disease whether structural or functional—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 19th century. The ancients even maintained that the heart was not subject to disease—*cor non aegrotare posse*, as Hippocrates is said to put it.† Galen's classification of types of heart disease (wounds and inflammations, pericarditis and pericardial effusions, palpitations) disregarded any logical sequence and, as might be expected, was woefully incomplete

* In the short space available, it is obvious that frequently the bare mention of a name must suffice. Fortunately, the details of important contributions—often, the pertinent portions of the original texts, in English translation—by such leaders as Benivieni, Haller, Morgagni, Vieussens, Senac, Heberden, Parry, Adams, Stokes, Cowper, Duroziez, Potain, Corrigan, Hope, W. His, Jr., Keith and Flack, can be found in Willis and Keys' *Cardiac Classics*, Major's *Classic Descriptions of Disease*, and several volumes of Kelly's *Medical Classics*, and other anthologies.

† This statement is offered by both Moon and Herrick as the basis for the erroneous belief 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.

to modern eyes. Yet it controlled the current of medical thought for some 1300 years. With very few exceptions, it was only in the 15th century that even gross anatomic changes began to be recorded. Pietro di Montagnana (died 1460), for instance, noted damaged hearts in 14 dissections at Padua.

Functional disturbances such as palpitation (said to be frequently mentioned by Hippocrates) and arrhythmia (a term attributed to Galen) were naturally recognized much earlier though the word "palpitation" until recent times covered a wide area ranging from a violent pulsation to an increased activity due to diseases. An irregular pulse meant little more than an irregular pulse until instruments of precision led to classification of the various arrhythmias into objectively recognizable types. These are considered in Chapter V, B and their story outlined later in this chapter. The two branches of the subject, structural and functional, were, to be sure, often mingled in what today would be called a clinicopathologic study, straight pathologic anatomic writing gradually emerging as knowledge of this branch became more extensive.

The development of knowledge of pathologic changes in the heart came slowly, as it did for most of the internal organs. This growth can conveniently be considered in three overlapping periods: the first, the longest and least valuable today, characterized by

140 T. H. KERCKRINGII

Tabula xxii. ad Observationem Lxi. cor triplici ventriculo præditum ostendit.

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

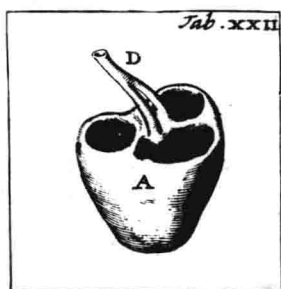


Figure I-1. From Kerckring's report (in *Spicilegium Anatomicum*, 1670, Obs. 69, Table 22), showing a heart with a third ventricle. D, Pulmonary artery arising from the double right ventricle. (The letters BB and C do not appear in the original illustration.) Note the crudity characteristic of many 17th century illustrations.

isolated observations of such material as chance offered; the second, in the 17th and 18th centuries, a period of systematized collections of cases usually including some clinical material; and the third, in the 19th and 20th centuries, a period of treatises and textbooks, with increasing attention to the laws underlying the observations. Almost negligible through ancient, classical and medieval times, for practical purposes the beginning of cardiac pathology may conveniently be located in the Renaissance with Benivieni's *De Abditis Morborum Causis* (published posthumously, 1507). Of the 111 chapters in this small book, one, of the 20 that included a postmortem report, describes what seems to be an acute pericarditis, though it is not clear whether by "the cavity" *cum pilis refertum* (covered with hairs) is meant that of the pericardium or of one of the cardiac chambers. Another case revealed a "polyp of the heart," an erroneous

diagnosis that was not finally corrected until the 19th century (see Gross, 1857). However, disorders of the heart contributed through the centuries, though not as often as one would expect, to the progress of the anatomic concept of disease. Even in the great *Sepulchretum* (1679) of Bonetus, we find few significant items of cardiac pathology, though there are numerous cases of palpitation and cardiac polyps, some of the latter having been so diagnosed during life. These cardiac polyps had a history far out of proportion to their importance. Bauhin (1592), Tulp (1641), famed for his Rembrandt's Lesson in Anatomy, and Malpighi (*De Polypo Cordis*, 1686) were others who prolonged the confusion between ante- and postmortem clots, until Kerckring (1640-1693) showed that the red, easily removable kind was merely a postmortem clot (*Spicilegium Anatomicum*, Obs. 73, 1670). The error was gradually eliminated as the gray, tightly adherent ante-mortem thrombus became differentiated from the loose red variety. Even in 1839, in the first edition of Gross' well known *Elements of Pathological Anat-*



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

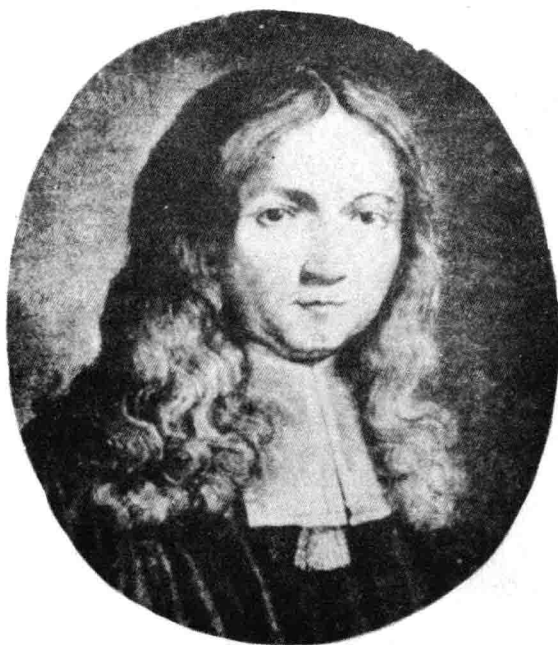


Figure I-3. Raymond Vieussens (1641-1715). (Reproduced by courtesy of Oxford University Press.)

omy, polyps appeared as "polypous concretions," though in the third edition (1857) "polypous" was changed to "fibrinous." Kerckring also was an early contributor to the pathology of congenital heart disease; his 69th observation showed an infant's heart with a large double right ventricle (*cor triplici ventriculo*) and a pulmonary artery leading from each, to fuse later, as is clearly shown in Figure I-1.

In the 17th century the custom of publishing assembled cases, from the literature as well as from personal observation, reached its highest development in Bonetus' *Sepulchretum* and then in Morgagni's great *De Sedibus et Causis Morborum* (1761), which is properly credited with having generally established the anatomic concept of disease. The *Sepulchretum*, though it included 2934 observations (cases) made by 470 authors, reports very little about the heart. Even Morgagni's *De Sedibus* contains much less about the heart than about other organs and systems. However, we find reports of the structural changes in a case of what was soon to be called angina pectoris; one of the earliest accounts of ap-

parent heart block (slow pulse with syncopal attacks); cases of vegetative endocarditis—one associated with gonorrhea; a case of rupture of the heart; one of myocardial degeneration; and one of congenital hypoplasia of the aorta,—other names of course being applied to most of these. Morgagni (Figure I-2) had the great merit of correlating careful clinical study with necropsy findings better than any of his predecessors and, for that matter, than many of his successors. One of the greatest medical figures of the 18th century, he deserves a prominent position in the history of the pathology of the heart.

Systematic descriptions of cardiac pathology, though of course not in any way comparable to those in modern texts, had occasionally appeared between the time of Benivieni and that of Morgagni. We think of four, all by prominent physicians—Fernel, Vieussens, Lancisi, Senac. Jean Fernel (1497-1558), a humanist who pursued all branches of human knowledge, conceived of a *Universa Medicina* which he never finished. In one of the three completed sections, *Pathologia*, he grouped heart diseases under the peculiar headings of inflammation, erysipelas, *tumores contra naturam*, ulcers and wounds. Over a century later, Raymond Vieussens (Figure I-3) produced his *Traité Nouveau de la Structure et des Causes des Mouvements du Coeur* (1715),* which has been called "the first to make serious contributions to our knowledge of diseases of the heart" (Moon). In addition to good anatomic descriptions of such items as the course of the coronary vessels and the valve in the coronary sinus, he described mitral stenosis (see Figure I-4) and the characteristic pulse of aortic regurgitation. (See also under Endocardium, page 17.) His contemporary, Giovanni Maria Lancisi (1654-1720; Figure I-5), for 13 years a Professor of Anatomy, while practicing all the while, became the greatest clinician in Italy. Important contributions to cardiovascular knowledge are to be found in both his celebrated books, *De Subitaneis Mortibus* (1707), and *De Motu*

* Published the year before his death, this text is extremely rare. In the catalogues of several great medical libraries it is only to be found concealed as Part 1 of his *Oeuvres Françaises* (1715).

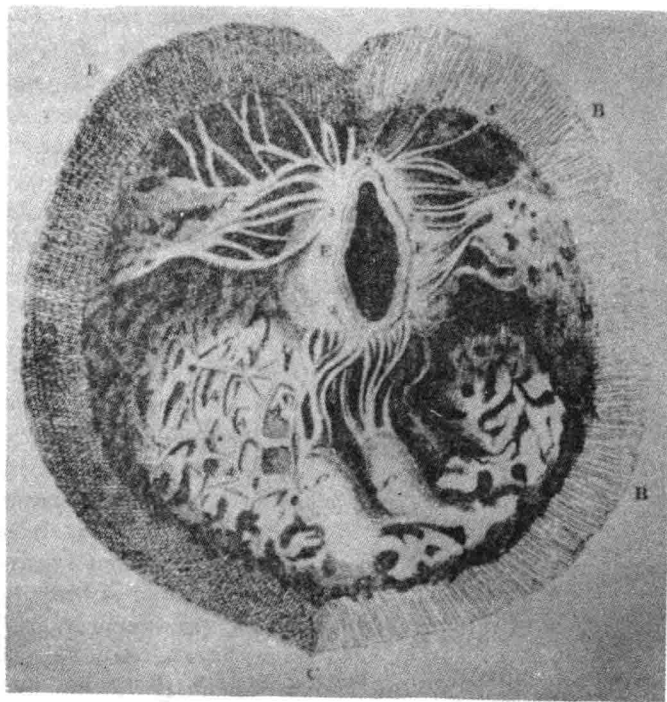


Figure I-4. Vieussens' illustration of a stenosed mitral valve from his *Traité Nouveau de la Structure et des Causes du Mouvement Naturel du Coeur* (Toulouse, 1715). (From Major's *History of Medicine*, courtesy of Charles C Thomas.)

Cordis et Aneurysmatibus (published posthumously, 1728). He recognized heart disease as one of the common causes of sudden death and gave good descriptions of sclerotic and of warty valves and of the coronary system. (See Figure I-6.) He also described some arteries that were furnished with narrower orifices (*angustioribus orificiis praediti sunt*). He dwelt at some length on cardiac "aneurysms" (in the strict etymologic sense of dilatation, a meaning still used for some aneurysms). Hence for him, aneurysm was commoner in the atrium than in the ventricle and least common in the left ventricle. His handling of the causes of heart disease in general was less successful, stressing congenital defects, violent emotions, physical effort, palpitations. The causes of cardiac hypertrophy hit somewhat nearer the mark: heredity, leaky valves, calcified arteries and valves, chronic asthma, true and false and syphilitic aneurysms. His contemporary, I. F. Albertini (1662-1746) favored heredity and syphilis (using mercury in its

treatment) as major causes of heart disease, and correlated dyspnea and pulmonary edema with it.

De la Structure du Coeur, de son Action et ses Maladies (1749) by Jean Baptiste Senac (1693-1770; Figure I-7) has been generally accepted as the most important extensive early work devoted entirely to the heart. In the chapters on the diseases of the heart he recognized inflammation of all three of its layers, and noted that pericarditis might follow pneumonia or pleurisy or infectious fevers; he also observed pulsation of the arteries of the neck when the left ventricle was enlarged and pulsation of the cervical veins when the right ventricle was enlarged (Major, *History of Medicine*, 1954, p. 633). Ossified coronaries and insufficient valves were other valuable observations. He included discussion of tumors, abscesses, "ulcers" and wounds, as well as arrhythmia, palpitation and syncope (weak action) of the heart. However, many of his descriptions, like those of his contemporaries,



Figure I-5. Giovanni Maria Lancisi (1654-1720). From an engraving by Sebastian Conca in the first edition of his *De Motu Cordis et Aneurysmatibus*, 1728. (Courtesy of Armed Forces Medical Library.)

are found in an atmosphere of such fanciful concepts that they make no great impression on the modern reader.

Textbooks of pathology, in the sense in which we use the word "textbook" today, may be said to have started modestly with Matthew Baillie's (1761-1823) 52-page work, *The Morbid Anatomy of Some of the Most Important Parts of the Human Body* (1793). In the 24 pages of the chapters on the heart and pericardium, he touches briefly on inflammation, abscess, gangrene, polyp, cardiac aneurysm, fibrous, bony, and "earthy" thickening of the valves, rupture of valves and myocardium, malformations and hypertrophy of the heart; and on "white spots," inflammation, adhesions, dropsy, excessive dryness, and scrofulous tumors of the pericardium. Clinical notes were included in the first edition only in the German translation. The early textbooks in France and Germany, such as Lobstein's *Traité d'Anatomie Pathologique* (1829) and the younger Johann Friedrich Meckel's *Hand-*

buch (1812, 1818) dealt with general pathology only, the special pathology being covered by atlases. On the other hand, the first textbook of any size in this country, *Elements of Pathological Anatomy* (1839), by Samuel D. Gross (1805-1884), was divided, as most teaching texts are today, into general and specialized pathologic anatomy. But in the latter portion, the heart and its membranes occupied only 40 of the 510 pages. Rupture of the heart was regarded by Gross, and for most of a century after, as "generally the result of ulceration or of the softening of fatty degeneration."

The French School, dominant in the early 19th century because of the political situation in Europe and the French achievements in physical diagnosis, also set the pattern in the expanding knowledge of the pathology of the heart. (See Figure I-8.) Two great clinicians, Jean Nicholas Corvisart (Figure I-9) and R. T. H. Laennec (Figure I-10), led this advance. Corvisart's *Essai sur les Maladies et les Lésions Organiques du Coeur* (1806), by means of clinical lectures with a pathologic

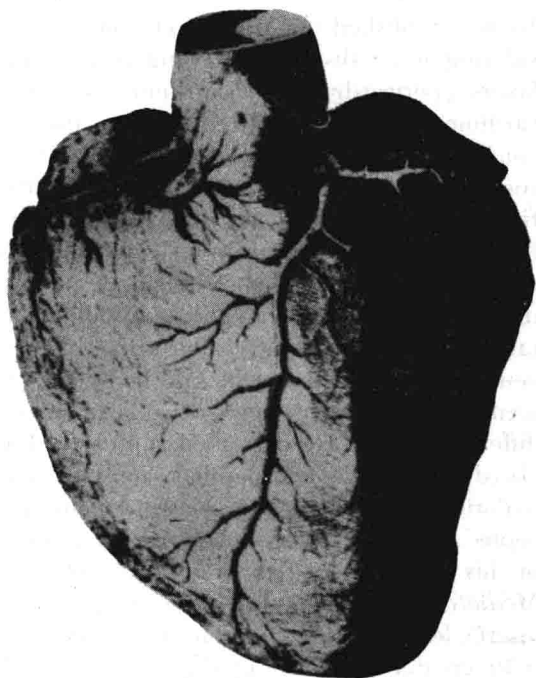


Figure I-6. Vieussens' illustration of the coronary arteries from the *Traité Nouveau*, Plate 5, anterior view. (Courtesy of Armed Forces Medical Library.)