

RHEUMATIC HEART DISEASE

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WITH AN INTRODUCTION BY

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*WITH NUMEROUS ORIGINAL PLATES
AND ILLUSTRATIONS IN THE TEXT.*

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PREFACE.

IT is a remarkable fact that so familiar a feature of everyday practice as rheumatic heart disease has not hitherto been made the subject of a monograph. In attempting to fill this gap I had to choose between one of two plans—either to compile a more or less critical review of the work of others, or to undertake the task of surveying the disease for myself. The latter plan was chosen; first, because it gives a greater unity and perhaps a stronger flavour of authority to a book if it is the offspring of one mind; and secondly, because the task of collecting and reviewing everything that has been written on the subject of cardiac rheumatism is beyond the power of any one man to accomplish.

Yet it is by no means true to claim that this book is the work of one mind. Indeed, it is doubtful whether the mind of the author had much to do with the writing of the book, as a brief historical survey of the work which has led up to its publication will show.

It began in 1903, when I was medical registrar at St. Mary's Hospital, in a conversation with Dr. Poynton, who suggested that the rheumatism of childhood would prove a congenial study to a young physician who was looking for something to work at, and who, moreover, had been clinical clerk to Dr. W. B. Cheadle, and house physician to Dr. D. B. Lees.

Migration to Bristol in 1904, with a clinical appointment at the Children's Hospital and a pathological one at

the General Hospital, gave both impetus and opportunity to the study of cardiac rheumatism. At the same time the kindness of former colleagues, Dr. Reginald Miller and Dr. E. H. Kettle in particular, maintained a link with the work being done in London.

I acknowledge gratefully the unreserved kindness of my colleagues in Bristol, who have at all times made it easy to pursue these investigations. My fellow physicians, at the Bristol General Hospital, and Professor Walker Hall, Dr. G. Scott Williamson, and Dr. Geoffrey Hadfield, must in particular be thanked for their invaluable help and advice.

Just before the war, finding it difficult to carry on any kind of research in competition with the increasing demands of other work, I happened to say something of the kind to Sir Clifford Allbutt, and it was owing to his encouragement that application was made for a grant from the Medical Research Council. From that time until 1923, apart from the interruption caused by the war, the Council gave me generous support, without which the histological investigations could not have been completed, nor those embodied in the chapter on "Course and Prognosis" undertaken.

I have also to thank the Grants Committee of the University of Bristol Colston Research Society for help with an inquiry into the incidence of rheumatic heart disease in Bristol, and with other phases of the work, particularly with histological investigations into the basis of auricular failure. Thanks are also due to Dr. C. E. K. Herapath for help in the reviewing of a number of 'follow-up' cases, and for the loan of electrocardiograms

and polygraph tracings ; to Dr. Geoffrey Hadfield for the photomicrographs ; and to Miss D. Pillers for the line drawings. Miss S. G. Tribe has undertaken all the secretarial part of the work with an enthusiasm that has been very encouraging and helpful ; and the publishers have shown unflinching consideration and courtesy in applying their long experience to the production of the volume.

The catalogue of indebtednesses must end, as it began, with the name of Dr. Poynton, who has been good enough to write an introduction to the book.

It is not too much to hope, perhaps, that this contribution may be thought worthy of inclusion among those works of the St. Mary's school that, from the time of Sibson to the present day, have added so much to our knowledge of cardiac infections, and opened up a prospect of prevention of organic heart disease.

C. F. C.

BRISTOL,

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INTRODUCTION.

THERE are probably few more severe tests of 'optimism' than a survey of one of our well equipped medical libraries. The zealous investigator or student walks into it glowing with enthusiasm; but at the vision of tables piled with numberless periodicals, and shelves crammed with volumes upon every conceivable medical subject, his enthusiasm changes to dismay; the quiet somnolent atmosphere saps his fibre, and as the courteous librarian fills up his allotted table with the literature he seeks, he gently falls asleep. There is excuse for him, for in these days of stress and detail some new discovery of greater or less magnitude is continually appearing, and there follows inevitably an avalanche of new editions and critical essays.

Fortunately the human intelligence has not yet become so jaded as not to appreciate a book which has a clear lesson to give, particularly if the writer shows that he has learnt the lesson himself at first hand. In this volume, to which Dr. Carey Coombs has asked me to write a short introduction, the clear lesson is the far-reaching importance of rheumatic heart disease, and the pages are filled with personal observations and investigations. The request on his part was a kindly act, and when he gave as one reason, that he had derived some inspiration and encouragement from my own contributions to the subject, I felt it was also a special honour to accept this invitation.

Dr. Coombs' volume has been published at a most opportune time, for in this country and abroad there has been a rapidly growing realization of the gravity of rheumatic diseases, and it is my conviction that the

result of this movement will be of great practical importance. Acute rheumatism is the most important and frequent cause of organic heart disease, and if it can be prevented or minimized there will result the most promising advance in cardiac therapeutics of modern times.

The fact that this book contains many independent observations, and that these are utilized with the particular intent of forwarding prevention, contributes largely to its practical value. Another feature which particularly appeals to the writer is the thorough appreciation by Dr. Coombs of the importance of the study of a heart infection such as the rheumatic as an entity, and not as one of the varieties of a pericarditis, endocarditis, or myocarditis. In a small manual on heart disease written in 1907, I ventured in the preface to claim that this method of treatment of the subject was one which marked a step forward in principle. Although at that time the statement attracted little attention, the present volume appears to me to bear out its truth. If this method were generally adopted in text-books, far more convincing pictures of the various heart affections would be obtained than from descriptions of the different independent lesions. Whatever minor criticisms may be made of Dr. Coombs' monograph, no one can fail to recognize that a description such as is given of rheumatic heart disease at all ages gives us an insight into the complex problems of heart affections, and a sense of proportion of the various phases and dangers, which could be obtained by no other method of handling.

There is another advantage also in this method. All students of acute rheumatism recognize the variability in the manifestations from year to year. In some years subcutaneous nodules are unusually frequent, in others carditis, in others chorea. This variability is also noticed in different parts of the country, and we have in this book a description of the disease as it occurs in the Bristol area.

Dr. Coombs points out that he is under the impression that cardiac disease is more frequent among London children. This, so far as I can judge, is the case, and I believe that the disease is not only more frequent, but occurs at an earlier age. I also believe that aortic disease is commoner, and of recent years have been impressed with its frequency even in very young children. Holding the particular views I do, this interests me, because I look upon multiple valvular lesions as early signs of a malignancy in type, and have found that such are the cases which in adolescence or early adult life are more liable to develop frank malignant endocarditis.

The etiology of acute rheumatism will certainly be one of the problems that readers of this book will approach with interest, for there has been much speculation upon it, from the earliest writers on the subject up to the present time. A careful review is given and the present attitude is plainly indicated.

Dr. Coombs, as a result of his own independent observations and study, has clearly come to the conclusion that the most probable exciting cause is a minute streptodiplococcus. Throughout the various chapters we find that this cause is used as the most serviceable explanation of clinical and pathological facts. He neither hides the difficulties nor doubts that further investigations by the most modern methods and with the most recent technique are essential. To the writer it is naturally of great interest to find an authority on the subject of rheumatic heart disease making a bold use of his belief in the streptodiplococcus, not content to make a merely passing allusion to this theory only to dismiss it as unproven. Whether Dr. Coombs' faith is to be justified or not in the future, he has done a valuable service at this juncture in presenting an independent account of the disease based upon this theory of the etiology, and his knowledge of the questions involved makes his contribution one of special importance.

Many years ago Dr. Paine and the writer foretold that the real struggle over the etiology would be fought, not around acute rheumatism and its heart disease, but around malignant endocarditis. Time has proved the correctness of this prophesy, for with the evolution of 'subacute bacterial endocarditis' the storm centre has moved in this direction. It was not difficult to foresee this, for it is comparatively easy to study the bacteriology of malignant endocarditis, but difficult to study the bacteriology of acute rheumatic heart disease, because in the majority of cases the latter is not fatal, and when it is, often enough the bacteria have been destroyed and we are left with the lethal results of the toxins. The relation of malignant endocarditis to simple endocarditis is very wisely left an open question by the author, for the time has not yet come for a full decision upon this point.

The many secondary factors in the causation of acute rheumatism are fully considered, and the directions in which search should be made for further data are discussed; this is very essential, for the problem of acute rheumatism is concerned with an infection which in all probability is greatly influenced both by secondary extrinsic factors and also by the intrinsic factors of human resistance.

In the chapters upon the morbid anatomy and physiology there is much independent observation and research, and particular attention is given to the Aschoff bodies or nodes. These lesions have attracted many investigators since Aschoff first described them in detail.

In 1899 the writer demonstrated the local lesions in the myocardium at a meeting of the Royal Medico-Chirurgical Society, but neither he nor Dr. Paine had attached the specific importance to them later ascribed by Aschoff. At the present time, while entirely in agreement with the value of the numerous investigations that have been made, he is not prepared to see in these nodes more than a phase

of a healing lesion. Such lesions are frequent in rheumatic conditions because rheumatic injuries tend to heal, but it remains in his opinion doubtful whether these nodes are specific except in this sense, and he would not be prepared to look upon their presence as a necessary proof of the rheumatic infection. The specific pathological lesion is a conception which in human disease requires very cautious acceptance.

Dr. Coombs makes some interesting observations upon the *a-v* bundle, pointing out that there is no special incidence of the rheumatic inflammation upon this structure. This observation bears out his clinical experience that disturbances of rhythm in childhood are not a striking feature of rheumatic heart disease, which has also been the experience of the writer.

We both also find ourselves in agreement over the rarity of extensive pericardial effusion in rheumatic pericarditis under twelve years of age, and the practical importance of this is clearly pointed out.

The valuable papers of the late Dr. D. B. Lees upon acute dilatation are recognized, and their application to the study of acute carditis confirmed and strengthened by further observations.

The chapters upon the symptoms and physical signs of rheumatic heart disease are full of interesting details, and illustrate very distinctly that the study of cardiac rheumatism makes an excellent introduction to the study of cardiac disease in general, and confirms my belief that the best early training for any one who wishes to make this branch of medicine his particular line is to be found in the wards of a children's hospital. Here we can trace the early stages of mitral stenosis and aortic regurgitation, and appreciate the importance of myocardial disease, uncomplicated by the secondary factors of unsuitable occupations, alcoholism, and faulty habits.

Dr. Coombs insists on the danger of overlooking primary cardiac rheumatism, which of all forms of rheumatism in

the young is the most insidious, and which, in any scheme for the prevention of the disease, is the manifestation which will give us the greatest difficulty. In such cases the arthritic and muscular pains may be slight, easily overlooked by busy and not too observant parents; and every doctor recognizes that the actual symptoms of early heart disease in the young may be negative until they have advanced far enough to cause obvious dyspnoea or possibly pain.

Dyspnoea, which is classified as acute, persistent, and paroxysmal, leads the author to dwell upon the ill effects of heavy work on the damaged heart, of which he gives illustrations. No one can doubt that it is our duty to exert an ever increasing vigilance in the prevention of such calamities, which may ruin an otherwise useful life, and rapidly undo all the care and skill, to say nothing of the expense, which had been previously devoted to shepherding the individual through the original attack of cardiac rheumatism.

In the study of rheumatic heart disease it is essential that we keep before us the fact that the great cause of a cardiac breakdown in the young is a fresh or recrudescent infection.

When there are mechanical difficulties from the result of scar formation caused by previous attacks, it may be difficult at first sight to recognize the active infection which is sapping the power of the heart. The author brings out very clearly the importance of this factor, which was first apparent to the writer in an analysis he made in 1898, when in 100 fatal cases in children under twelve years of age he found signs of activity of the infection in 86 per cent. Throughout the life of a patient suffering from rheumatic heart disease this element is likely to appear, although with increasing years the mechanical disabilities of the heart become progressively in the ascendant.

The difficult question of pain in rheumatic heart disease

meets with due attention, and reference is made to the puzzling and obscure acute attacks that sometimes occur.

In an interesting chapter on disturbances of rhythm, Dr. Coombs gives some interesting observations upon the age incidence of auricular fibrillation, and his experience of arrhythmia in the young is a useful contribution to a subject which is not easily investigated except under the most favourable circumstances.

The chapters on diagnosis, prognosis, and treatment lead up to the most important practical consideration in the book, the possibility of prevention, and they are on this account of special interest. It is from this point of view that the difficulties in diagnosis are of chief importance. Accuracy in this respect is undoubtedly a valuable asset, but from the aspect of prevention it is of minor importance if a case of tuberculous or pneumococcal heart disease is mistaken for the rheumatic, because both are in comparison rare, and little or no harm can result from such an error; on the other hand, to overlook a heart affection from lack of examination is serious, and the need for careful investigation in all doubtful or likely cases is commented upon by Dr. Coombs. Equally important is the danger of condemning a child's heart as organically damaged when there is no actual disease, and this is a very real pitfall. There are cases in which it may be most difficult at a first examination to come to a decision, and we cannot be too careful in reviewing every scrap of evidence before committing ourselves to an organic lesion. In any scheme for prevention there must always be a danger of disagreement, and there will undoubtedly be as much difficulty upon this point as there is at present over the diagnosis of early pulmonary tuberculosis in childhood.

The chapter upon the prognosis of rheumatic heart disease is a very important contribution, and contains observations of particular value. There is general agreement upon the gravity and frequency of this condition,

and there is also a general recognition that in a considerable number of cases remarkable improvement occurs.

Here we have some data which enable us to arrive at accurate conclusions. By the aid of a grant from the Medical Research Council Dr. Coombs was enabled to place in the hands of Mr. W. S. Emery the notes of over 600 cases for the purpose of following up their subsequent histories. Anyone who has attempted even on a small scale such an inquiry can realize the time and energy required for such a search, and the frequent disappointments that are met with.

The results are placed in two divisions, one containing the patients who were first seen in the stage of active heart disease, the other those who had already developed lesions associated with the adult stage. In the first 374 cases were traced, in the second 281, a remarkable achievement! In the first group, 253 were cases of undoubted active rheumatic heart disease, and 121 were suspicious cases. Here allusion will only be made to some of the important generalizations that Dr. Coombs has made.

Firstly, with regard to the group of undoubted cases, he found that about one-twentieth died within twelve months. Secondly, of the suspicious and doubtful cases, one-third within ten to sixteen years showed definite signs of valvular disease. This is a forcible illustration of the need for periodic cardiac examinations in suspicious cases.

Another interesting result from the following-up of 200 cases of undoubted early rheumatic heart disease was the discovery that a quarter of them had lost all physical signs in fifteen years. This is valuable evidence in support of the view that cases which may show clear signs of organic heart affection may recover so thoroughly as to defy detection. This is in entire accord with the writer's experience. He has supervised children who at the ages of six or seven years have had most definite attacks of carditis with dilatation and mitral systolic murmurs heard over a wide area round the left axilla to

the back ; at that time neither he nor the family doctor has had the slightest doubt as to the condition ; and yet when the age has come to go to a preparatory school the school doctor has thought this diagnosis was a mistaken one, recovery had been so complete. Such facts as these are very comforting to those who believe in the future of prevention.

Turning to the second group of 281 cases with established heart disease when first under observation, 83 were traced to have survived the age of forty years.

Another very interesting series of observations in this chapter is concerned with the effect of rheumatic heart disease upon the working capacity of the individual. This is clearly a question of the most far-reaching importance, not only from the standpoint of the individual, but also because when such investigations have been accumulated and analysis made on a grand scale, we shall have a definite and not a nebulous idea of the national wastage from this disease. Further, if measures for prevention can be eventually organized, also on a grand scale, another series of analyses in the future will enable us to gain some definite conception of their value by comparison with those made before such measures were undertaken. In his chapter upon treatment Dr. Coombs very wisely lays stress upon the time that must elapse before we can expect to elaborate and test any efforts for prevention : any idea that such a difficult problem is to be solved hastily is most certainly one that cannot be entertained. The question of efficiency and rheumatic heart disease is one of extreme interest, and its study must produce most valuable results in the future.

In the final chapter, many of the points bearing upon prevention are dealt with. Foremost stands the convalescent hospital, the most urgent but unfortunately the most costly requirement. There can be no doubt that this need will gradually be supplied ; not, the writer hopes, necessarily in the form of special cardiac hospitals,

but as cardiac sections in the general ones. Looking into the future, one foresees the gradual shifting of most of the sick, except urgencies and out-patients, from the expensive and inappropriate surroundings of a large town.

The need for further research in the laboratories, and also afield into the influences of toil, overcrowding, and so on, come next. Then follow the importance of the education of the students of medicine and of the public in the dangers of rheumatic heart disease, and the influence of educational strain. Lastly the use and abuse of passive rest and the immense value of occupation are considered.

Dr. Coombs inclines to the value of local investigations in restricted areas, and these certainly promise valuable results. The writer believes that what he would term official English medicine will eventually afford the greatest assistance, and hopes that in the pursuit of the common aim there will develop that increasing co-operation between official and voluntary effort which no one can doubt is essential for the welfare of the health of this country.

The author does good service in dwelling upon the great value of occupation for the victims of rheumatic morbus cordis. This is at present a difficulty, not only in adult life, but also in childhood. The partially invalid child, staying at home with parents too busy or too negligent to occupy him, drifts often enough into an aimless and depressing atmosphere which is so injurious that one is forced to believe a return to school life, even if premature, may be the lesser of the two evils.

In concluding this introduction, the writer repeats that this volume has come at a most opportune time. The medical profession in the last twenty years has in the face of great difficulties made great efforts in preventive medicine. Tuberculosis and venereal diseases have been taken in hand; cancer research, although not yet rewarded by substantial success, is ready to pursue any new light that may appear, though it may be from some quite unexpected quarter; infant welfare and the

activities of the school medical services are pressing forward. Now surely the time has come to turn to 'rheumatism', and in particular to acute rheumatism in the young! Such patients must have the sympathy of all, for none appeals more than the child with rheumatic heart disease, who is the most alert and engaging of all our patients. If ever a medical venture on a large scale was indicated in this country, it is the prevention of rheumatic heart disease, and by this the writer means the supervision and care of the rheumatic child, adolescent, and adult.

Dr. Coombs' monograph puts before us the problem with which we have to deal in plain terms, and, more important still, justifies our confidence that by prevention great good will ultimately result in those damaged by rheumatism, and that greater knowledge will be obtained of heart disease in general.

F. JOHN POYNTON.

CONTENTS.

CHAPTER	PAGE
I.—HISTOLOGICAL AND INTRODUCTORY	I
II.—ETIOLOGY	5
III.—MORBID ANATOMY AND HISTOLOGY	43
IV.—MORBID PHYSIOLOGY	78
V.—SYMPTOMS	121
VI.—PHYSICAL SIGNS	159
VII.—THE PULSE : ARRHYTHMIA	209
VIII.—DIAGNOSIS	233
IX.—COURSE AND PROGNOSIS	264
X.—TREATMENT	316
XI.—PREVENTION	348

Rheumatic Heart Disease.

CHAPTER I.

HISTORICAL AND INTRODUCTORY.

THE history of our knowledge of rheumatic heart disease does not differ from that of most scientific observations. First of all there is a period of twilight, than a few plain facts emerge. Soon these are grouped together in some sort of law and order until a concrete picture of the whole subject can be painted, so far completely, at all events, as to define not only that which we do know, but also that which we do not yet know, and must therefore study. The statement which this book professes to make has both ends in view. It is quite time that so important a matter should receive some more systematic attention than has hitherto fallen to its lot. To this end I propose to write down as well as may be, a summary of studies which have occupied over twenty years, both for the help of those who would like to know how the matter stands at the present day, and also perhaps for the guidance of some who would like to investigate all the as yet unexplored areas of this field of medicine.

Lancisi¹ refers to an autopsy made on Sept. 26, 1706, at which fleshy swellings of the aortic valves were noted. These are so clearly described that there is no doubt as to their inflammatory origin, but they were of the 'malignant' rather than of the 'rheumatic' type.

In the works of Morgagni² there is a plain allusion to