Course and prognosis of rheumatic heart disease

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COURSE AND PROGNOSIS OF RHUMLATIC HEART DISEASE

Wm. F. Logan

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INTRODUCTION

Rheumatic heart disease is that disturbance of the heart resulting from the same etiological agent that produces the rheumatic state. It is becoming more evident that the cardiac involvement is as much a manifestation of this disease as the other symptoms, such as rheumatism and chorea. For some time the term rheumatic heart disease was questioned, since many gave no definite history of antecedent rheumatic infection. However, to insist upon a preceding polyarthritis or chorea before one makes a diagnosis of rheumatic heart disease goes back to the obsolete conception of heart disease as a complication of acute rheumatism in childhood, rather than an integral part of the rheumatic process. The cardiac manifestations of the rheumatic state are by far the most important, being not only the most common but the chief scourge of childhood and young adults. Realization of this fact has stimulated great interest in this problem.

That this disease extends from its often stormy febrile beginning to an end having apparently little relation to it, required the observation of many decades to suggest. But this fact is now definitely established. However, what is least understood is the fate of the individual in the intervening period. It is one purpose of this paper to trace the course of this disease from the beginning to the end with its many side branches. Another, is to try to outline both the immediate and ultimate prognosis at each stage of the disease. For after all, one of the main things the patient wants to know is what is going to happen to him now and what he can expect in the future.

For the purpose of discussion, I have rather arbitrarily divided this paper into four main divisions. First, I have concerned myself
with the acute or active phase of the disease which is the first clinical manifestation usually observed. Secondly, is the phase that commands less attention and which, following Levine's example, I have chosen to label "potential heart disease." The discussion of chronic valvular heart disease of rheumatic origin constitutes the third division. In the fourth, I have grouped together statistical reports concerning the duration and termination of this disease.
ACUTE HEART INVOLVEMENT

It is extremely difficult to estimate the incidence of heart involvement in the rheumatic state. Many factors must be considered; first it is necessary to define just what is meant by heart involvement, next it appears that the heart is involved in mild cases of rheumatic fever without presenting any evidence of its involvement. If one utilizes all available means to detect any abnormality in the heart, it appears that approximately 100% show evidence of heart damage. (39)

It is true that many of these are slight and transient, some may be permanent and still not produce any discomfort to the patient. However, in some cases during the early stages of the disease, there is no demonstrable evidence of cardiac damage. (72) Some authors have estimated what they believed to be the incidence, but in considering these figures one must take note of the difficulty in arriving at their deductions. (71) Coombs gave an estimate of 72.5%; Dr. Sibson in 325 cases of rheumatic infection considered that only 79 cases had escaped cardiac involvement; Church divided his cases into two age groups and found that 75% under the age of 20 years and 57.5% of all ages developed heart damage from rheumatic infection. Regardless of how accurate these figures may be, one can easily deduct that the incidence of cardiac involvement in rheumatic patients is extremely high. The advent of the electrocardiogram has furnished us with evidence that there is some degree of functional disturbance in over 90% of the patients suffering with this disease. A valuable conclusion to draw, is that the heart is probably affected to some degree in practically every patient unfortunate enough to have an attack of rheumatic fever. Also we are able to assume that the first attack of rheumatic infection represents the time of onset of
rheumatic heart disease. However, it must be remembered that many primary manifestations of rheumatic fever go unrecognized and the discovery of cardiac damage, months or years later, may be erroneously considered the time of onset. (72)

AGE OF ONSET - While has recorded instances occurring in nursing infants. (72) However, such an early onset is very rarely seen. Davis and Weiss (18) list their earliest case at 3 years while De Graff and Lingg (20) found their earliest case at 2 years. But instances of the first attack are seen even at the age of 63 years and one must consider that at no age is one altogether exempt from a first attack. (43) The incidence of rheumatic fever shows a smooth regular curve starting with 4.7% under the age of 5; rising to a peak of 30.8% between 10 to 15 years, followed by a slow fall to 1.5% between the ages of 25 and 55. There is a rise in later years due to a confusion with other arthritides and because of the small number of cases. (43)

The average age of onset as noted ranges from the age of 7 to 14.6 while the mode, or year of greatest incidence occurs around the 7th or 8th year. Coombs (14) gives the average age of onset at 10.2 years in a young group and 14.6 in an older group. Poyton (51) gives the average at 7.0 years; Wilson (81) and Folk (65) 7.3 years; Ash (2) at 6.8 years; and Stroud and Twaddle (66) an average of 8.0 years.

In a series of cases studied by Lingg and Croxford (81) 50% occurred between the ages of 6 and 9, while Mackies (80) had only 23.9% occurring between the same ages. The majority occurred at a higher age. This difference is accounted for by the fact that the latter series included adults while the former age incidence was under 22. De Graff and Lingg (20) found that 84.5% were infected before 30 while
only 4.4% after 40. Willius (78) found very similar figures. In his series, 85% occurred before the age of 30 and 64% before 20 years while in only 3% did the primary infection occur after the 40th year.

From the preceding figures, it can be seen how impossible it would be to set any age as having the highest incidence of first attacks. Therefore, the age of onset is usually regarded in a broad sense as representing an age group. Most authors refer to the span of 5 to 15 years as having the greatest incidence of first attacks, while others narrow it down even to between 7 and 12 years of age. It is equally important to realize that no age is entirely exempt.

INFLUENCE OF AGE OF ONSET - It is generally thought that the younger the child at the time of the initial attack of rheumatic fever the worse his prognosis will be. (28) However, Coombs (14) found that there is not as much influence as usually believed. Using as a criteria, the fatalities within 10 years observation and the duration of these cases, he found little difference between those with the initial attack in the first decade and those in which it occurred in the second. Bland and Jones (5) are also in accord with this observation. In a study of 250 children that died with rheumatic heart disease they found no significant difference between the age of onset of those patients who succumbed to rheumatic infection within one year and the age of onset of those who died after a longer period of time.

<table>
<thead>
<tr>
<th>Duration in years</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
<th>8</th>
<th>9</th>
<th>10</th>
<th>11-15</th>
<th>16--</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ave. age onset</td>
<td>8.2</td>
<td>8.1</td>
<td>7.5</td>
<td>7</td>
<td>8.7</td>
<td>8.1</td>
<td>7.6</td>
<td>7.6</td>
<td>8.1</td>
<td>8.0</td>
<td>8</td>
<td>7.5</td>
</tr>
</tbody>
</table>
It was also surprising to find that the course was definitely shorter when the onset occurred after the 20th year than if it began in childhood or adolescence. In such cases some believe the onset was much earlier than this and that the attack after 20 years was in reality a recurrence. (14) How true this is is difficult to determine. It is true, however, that the disease runs a rapid fatal course more often in childhood, but this can be explained as due to the fact that the majority of cases occur in childhood (14). The apparent conclusion is that the age of onset does not greatly influence the number of years that elapse before death.

INFLUENCE OF TYPE OF ONSET - The type of onset definitely does influence the duration of the disease and its severity. Briefly, of the four great manifestations of rheumatic infection that with the most serious omen is the cutaneous nodules. Approximately one half of the patients presenting this symptom die within the first decade following their appearance. (14) The association of nodules with carditis is common knowledge. However, the presence of these nodules indicates only an extensive and severe infection and therefore the cardiac damage is often severe. (57) Carditis is the next most important manifestation and of these patients over one fourth die in the first decade. Of those who began with joint symptoms about one fifth failed to survive the same period of time. Chorea is by far the most benign form of the disease. In patients who presented this symptom as the initial attack, there were only one in thirty that died within the first decade. (14) Bland and Jones (30) in a study of 482 patients with chorea, over a period of 8 years from the onset, found that rheumatic heart disease developed in
and that there was only one death. Of 348 patients with chorea and other manifestations, 73% developed rheumatic heart disease and 14% died. While in 518 patients without chorea as a manifestation, 86% had cardiac involvement and the death rate was 32%. The relative mildness of chorea is quite evident from these findings.

**CARDITIS - MANIFESTATION OF HEART INVOLVEMENT**

During the acute rheumatic infection, it is frequently difficult to tell whether the heart is being affected and whether the changes noted are indicative of a transient or permanent cardiac condition. The incidence of cardiac manifestations appears to differ with the age of the patient. Generally the older the patient the more marked are the articular lesions and the less marked the cardiac manifestations. In children, there are few attacked below the age of 16 that escape without damage to the heart. This is well shown in the following table.

<table>
<thead>
<tr>
<th>Age Period</th>
<th>Percentage Cardiac Damage</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-10 years</td>
<td>75%</td>
</tr>
<tr>
<td>10-19</td>
<td>54%</td>
</tr>
<tr>
<td>20-29</td>
<td>30.6%</td>
</tr>
<tr>
<td>30-39</td>
<td>33.3%</td>
</tr>
<tr>
<td>40--</td>
<td>12.5%</td>
</tr>
</tbody>
</table>

Percentage cardiac damage in age periods (34)

Mackie (44) also noted that the incidence of cardiac involvement in the initial attack varied with the age. They found that the incidence curve approximately parallels that of the age of onset of rheumatic fever. Starting with 61.5% in the group under 5 years, it reaches a peak between the ages of 5 and 15 when 78% were found to have evidence of cardiac complications. It was only after the age of 25 that more than one half of the patients escaped cardiac involvement. There is evi-
dence that the heart is practically always involved to some degree. However, due to the mildness of symptoms, the difficulty in recognizing the presence of an active carditis, and the regression of characteristic physical signs, some observers have concluded that in a certain percentage of rheumatic individuals the heart is not involved. (51) Even considering this factor there is quite definite evidence of a high incidence of carditis as a manifestation of rheumatic fever.

TACHYCARDIA - Tachycardia need not indicate that the heart is seriously damaged or that it will show structural changes after the infection is over. A rapid pulse is perhaps one of the most frequent signs observed in the acute phase. However, it is necessary to view this sign with caution since many other factors than a carditis may be involved. A true tachycardia resulting from slightly damaged heart is very constant, a feature which should be observed before suspecting any cardiac involvement. (39) Poynton (52) was struck with the frequency with which mitral stenosis eventually developed in patients who presented tachycardia during the acute phase.

MURMURS - The most common sign of valvular disturbance is the apical systolic murmur of mitral insufficiency. This murmur frequently begins as a blurring and lengthening of the first cardiac sound and later develops into a definite soft systolic murmur. (39) The appearance of these so called organic murmurs during the course of rheumatic fever is generally accepted as evidence of valve injury. These are often the first clinical evidence of cardiac involvement. However, they are by far the most difficult feature to properly appraise during the acute infection. (4) Generally, it is an evidence of myocardial rather than
endocardial damage. There are two factors that may cause this murmur. First, it may be due to a local hyperemia set up in the valve producing temporary incontinence by weakening the valve structure. Second, the cause may be a myocarditis with a weakening of the cardiac muscle and consequent stretching of the chambers of the heart and the ring of insertion of the valves. (34) (72)

The knowledge that murmurs convey is of very great importance, but important as this knowledge is, it has certain limits which are not always sufficiently understood and as a consequence the practitioner is liable to fall into unfortunate errors of opinion as regards the gravity of the situation. In the past, the presence of a systolic murmur implied a serious diagnosis. But recently systolic murmurs have been found so often to be benign that an entirely opposite point of view has been developed. The correct evaluation is a serious though often a difficult task.

In adults, a systolic murmur has lost much of its significance. But in a child the discovery of such a murmur is a matter of some concern, particularly if it had not been heard previously. Experience has shown that the first rheumatic onslaught giving rise to a systolic murmur may be a stage in the development of a serious myocarditis or a progressive mitral stenosis. (57) This, of course raises the question as to how one is going to tell when the murmur is of significance. Unfortunately this cannot be answered definitely. However, Straud et al (65) have pointed out how important the intensity of the murmur is in indicating its significance. The louder the systolic murmur during the acute phase the more likely that it will be permanent and that it is indicative of some organic lesion. This is more particularly true if a
murmur of moderate intensity is present while the heart action is comparatively slow. Loud systolic murmurs rarely disappear, while faint systolic murmurs may either disappear, or if they persist may be practically disregarded. A systolic murmur will have more significance if; the heart rate is slow; there is no fever; no anemia; no hyperthyroidism and when the blood pressure is low. In the absence of these circumstances there is more likely to be some organic change. It is clear, that we must attempt to properly evaluate these murmurs because sometimes they represent a serious involvement of the heart. And when the condition runs an unfavorable course, symptoms of decompensation will develop. On the other hand, this murmur may be benign and of no significance whatever. Also, even murmurs which are due to myocardial involvement, usually decrease in intensity and some will entirely disappear following the subsidence of the acute phase. (52) (72) (42)

Diastolic murmurs are occasionally heard at the mitral area in early rheumatic carditis, especially in children. These are mid-diastolic in time and soft in character. It is rare that a murmur typical of mitral stenosis is observed in the acute phase. (34) Coombs (14) believes that these diastolic murmurs are always associated with ventricular dilatation especially of the left ventricle, which is responsible for the murmur rather than a valvular lesion. The evidence for such a conclusion is found at autopsy. Bland et all (3) is also in accord with the belief that the mid-diastolic murmurs at the apex, previously considered indicative of mitral stenosis are often due to dilatation of the musculature. However, Toussig (72) believes that they are due to actual involvement of the mitral valve. Schlessinger has termed it the murmur of potential mitral stenosis because while in some
cases mitral stenosis does develop, in other cases the murmur may entirely disappear. (57) However, Levine (39) believes that if evidence of stenosis is detected it is likely that the valve was previously injured and that one is not witnessing the first rheumatic infection but a recurrence. In any event the finding of a diastolic murmur is of more serious consequence than a systolic murmur and tho there is difference of opinion as to its significance, the patient demands careful attention and the prognosis regarded with caution.

Another murmur that may be found in the acute process is a diastolic murmur occurring along the left sternal border and indicative of aortic regurgitation. These murmurs are easily missed. Their significance lies in the fact that when the aortic valve is involved one is usually confronted with a severe type of infection, which may terminate in early life. These murmurs have been observed to disappear, so occasionally there may be recovery. (72) (52)

In a series of 1000 children with physical signs of valvular defect observed over a period of 10 years, Bland (4) and his associates noticed a regression of physical signs in a considerable number. In 83 cases there was complete disappearance of all clinical evidence of heart disease. In a few cardiac dilatation may have been responsible for the murmurs usually considered characteristic of valvular disease. If this is true, it is probably the explanation for the relatively rapid disappearance of the cardiac murmurs. However, they believe that in the majority of these 83 patients there was injury of the valve at the time of the acute phase but that it was of minimal degree in each instance seems most likely, so that the residual scarring has produced no significant deformity or characteristic murmur. In accord with a preceding view-
point, they found in no case did a very loud murmur disappear. It will be of considerable interest when these 83 patients have been followed and it is found out whether in any case there will be a reappearance of the murmurs, especially in those patients that will not have recurrence of the active infection.

CONDUCTION DISTURBANCES - Occasionally during the acute disease one finds a disturbance in the conduction of the cardiac impulses. There may be an actual block or an occasional omission of a entire heart cycle. This is indicative of an acute myocarditis which interferes with the conducting apparatus. Fortunately, recovery from such damage is very apt to be complete, the conduction disturbance tending to disappear. Even in rare instances where some permanent defect remains, the efficiency of the circulation may be perfectly normal. (39) Toussig (72) views them with a little more concern. He found that occasionally dropped beats occur but when they are numerous and detected with ease they are indicative of a severe infection. And when they occur early in the disease, they are liable to run a fulminating course with early fatal termination.

CARDIAC ENLARGEMENT - Cardiac enlargement may be the only cardiac lesion found in many cases of acute infection and it complicates all of the more severe rheumatic cardiac injuries. It is often the earliest and the most frequent evidence of rheumatic heart disease. (51) The diagnosis of this condition is very limited with only physical examination. In many cases it requires adequate X-ray studies. (82) Although during the acute stage it cannot be said that a heart that is normal in size is not diseased, it may be safely accepted that if the size of the heart increases or is greater than normal, it is diseased. (39) Marked cardiac
enlargement is of ominous prognostic import because primarily it is caused by myocardial injury. (72) Poynton (51) believes that the rapid development of acute dilatation accounts for some of the cases in childhood in which, after a slight arthritis, the patient has been allowed to return too quickly to ordinary life. And has suddenly become short of breath and collapsed with a syncopal attack. This serves as a warning to watch carefully the early days of what may seem a very mild attack. In rare cases it may prove fatal causing death by a clinical picture of asystoli. It is the chief cause of cardiac failure in recurrent attacks of carditis. Lastly, a large group in which the dilatation either is not severe and recovery is rapid or it may be slow in its course and intractable.

The subacute and chronic dilatations require more attention than they have received in childhood. Poynton (53) believes that they are an important cause of the so-called weak hearts in young adults. The usual course is that a child has a definite but not severe attack of acute rheumatism and the heart is found to be dilated with a possible mitral lesion. Instead of disappearing with recovery from the acute phase the heart remains enlarged, the murmur either disappearing or becoming more apparent. The child may be languid, short of breath and easily fatigued. When these individuals are able to go at their own pace and do a moderate amount of physical and mental work they appear quite normal. But if put under pressure, and particularly continuous pressure, their hearts become irritable and their nervous and digestive systems begin to fail. A very unsatisfactory condition may result in which nervous symptoms become predominant and in extreme cases may result in invalidism of the most trying kind, in bed they become quite happy and the heart symptoms
improve but a return to daily life causes a prompt relapse.

PERICARDITIS - In general, pericarditis is a mark of a severe infection and is repeatedly met with in fatal carditis. There is always a concomitant pancarditis. The presence of a pericardial friction rub is important mainly, because it indicates an extensive and severe infection. Massie and Levine (45) have estimated that pericarditis occurs in from 6 to 25% of the cases of acute rheumatic infection, while Ash (2) gives the incidence at 12.2%. The variations often found in the incidence could easily be due to the fact that the clinical diagnosis can easily be overlooked if the friction rub is not caught. Also, where the incidence is given by post mortem findings it is higher.(5)

On the whole, the presence of pericarditis indicates a rather poor prognosis. However, numerous patients after recovery from pericarditis are able eventually to lead a normal existence, and occasionally even lose all signs of previous infection. (2) The immediate mortality of 16.3% was found by Massie and Levine (45) The duration of the acute illness in these fatal cases varied from 1 to 18 weeks, with an average of 5 weeks. There was slight indication that the disease was more serious in the younger patients. All other factors seemed to be about the same in the fatal and non fatal groups.

In non fatal cases the acute attack may disappear in less than a week, the dilatation rapidly subsides and the patient will show a course of steady convalescence for about 3 weeks. Eventually the recovery may be complete. Far more frequently the active period is at least weeks in length. Later hypertrophy may develop and the signs of an adherent pericardium may slowly appear. The decompensation that follows is due to the myocardial disease rather than the adhesions. In the relapsing
form the symptoms may never be severe but the process advances at intervals of many weeks. The lesions do not completely heal and a fatal case may develop. (51) An attempt was made, in a study by Massie and Levine, (45) to investigate the role that pericarditis per se might play in the subsequent progress of those patients who recovered from the acute infection. His findings indicated that 83% of seventy cases did not show any progression while evidence developed in 18% of new valvular involvement or progression of a valve already involved. However, of the 18% three fourths had recurrences so that only about 4% could be said to have shown a natural progression of the original injury supposedly due to pericarditis.

AURICULAR FIBRILLATION - This arrhythmia rarely occurs in children the incidence being greatest between the age of 30 and 40 years. (48) (57) When it does occur in children those that have had frequent relapses and extensive cardiac damage are the ones selected. It seems to be a terminal event in children. (65) Schwartz (59) on the other hand, has emphasized the frequent occurrence of auricular fibrillation in children. However, he agrees that it is usually a terminal event. Because of its more frequent occurrence in the older group it will be discussed more in detail in a later section.

HEART FAILURE- When the acute cardiac infection is severe enough the cardiac reserve is spent and even minimal demands of a quiescent nature are too much for the heart and signs of decompensation appear. The cause of decompensation is not clear, but one is driven to the conclusion that the heart muscle is damaged by the rheumatic infection with lowered functional capacity not shown by histological changes. It is
essentially an activity of the rheumatic process having little if any
relation to mechanical strain. McKee points out that symptoms of decompensation so frequently follow recurrences that failure must be due to rheumatic process itself. With the appearance of symptoms the prognosis becomes accordingly grave. Edema appearing in a child, in the course of acute carditis, is a serious finding, the child is almost certainly to die within a few weeks. However, it is more by the degree of dyspnea that one is most often guided to an estimate of the degree of cardiac insufficiency, or rather by the amount of exertion that can be undertaken without causing discomfort. Cyanosis, visceral congestion and all the signs of decompensation are indicative of a poor prognosis. A sign of considerable prognostic value is fullness of the veins of the neck, in both the child and the adult. However, during the first attack only a small proportion of cases of rheumatic carditis are fatal. In some cases, even when decompensation is present it tends to clear up, tho such cases are rare.

GENERAL COURSE

Patients with acute rheumatic fever run quite a diversified course. The severity, duration and recurrences are so variable in different individuals that one can scarcely speak of a typical course. Yet, within these wide variations the disease runs remarkably true to form and experience teaches us to anticipate one of several clinical types. Brennemann (9) gives a very good division into clinical types. In the following presentation of clinical types I will follow his description closely.

FIRST GROUP - In the first group, is the child with acute rheumatic manifestations such as polyarthritis with fever and other symptoms of acute involvement. Upon examination of the heart, one will probably be impres-
sed with the absence of any positive findings. However, in some cases the examination may reveal a suspicious systolic murmur at the apex along with questionable cardiac enlargement. The temperature may return shortly to normal and the other signs of infection subside. If any cardiac findings were present, these will become less distinct and finally completely disappear. The child may reach adolescence without any recurrent attacks of rheumatic infection and to all intents and purpose be in good health without any symptoms or signs of cardiac damage and without any limitation in activity. Unfortunately this favorable course is unusual. More often the child, who has presented the picture given above, shows evidence of permanent valvular damage. Even though he may have no further attacks during childhood he is among the group who is likely to be handicapped later in life with evidence of chronic rheumatic heart disease.

SECOND GROUP - In the second clinical group, instead of a mild course of short duration, as seen in the previous picture, one is more likely to find that the symptoms and signs persist for many weeks or months. The fever persists or recurs, the appetite and general color is poor, there may be occasional pains in the joints or muscles, and some degree of dyspnea may be present. Together with this, there may be evidence of slight heart involvement. The liver may be enlarged, the veins of the neck engorged, and probably edema of the feet. After a variable period, these symptoms may improve, and finally disappear. The patient is more then likely to return to normal activity. In spite of the fact, that obvious signs of valvular deformity and heart enlargement sometimes appear, the patient may be entirely comfortable. After a variable period of quiescence covering either months or years, another episode of rheum-
atic infection usually occurs. This recurrence may be of the same nature as the first attack, or other manifestations of the rheumatic state may be apparent. And of greater importance is the fact that fresh evidence of cardiac invasion may appear. Regardless of the severity or the duration of these attacks, the heart practically always receives additional damage. Moreover, if the attack is severe enough he is likely to immerse with more cardiac enlargement and a greater diminution of the cardiac reserve. Repeated infections often lead to semi-invali- idism and the patient may even succumb before adolescence is reached. This second group, then, involves those with a more severe and prolonged infection with recurrences.

THIRD GROUP - Again the onset may be similar to the preceding group, but the symptoms and signs of active infection persist past the time that they disappeared in the other two groups. The patient may not even appear seriously ill during the early stage, but there is usually a low-grade fever, a pinched expression of the face, a rapid pulse and findings in the heart that give evidence of cardiac involvement. These cardiac signs are usually those of an apical systolic murmur and an increasing area of dullness consistent with enlargement of the heart. Perhaps in a few weeks the patient will show gradual improvement but only too soon there will appear exacerbations in his joints or even a friction rub of pericarditis. Often, subcutaneous nodules appear after such a course. During this time the child is usually dyspně and as the process becomes more extensive he develops orthopnea and other signs of congestive failure. Some patients succumb to the infection at this stage. If not, they may rally from these severe signs and then after a period of relative comfort, again become decompensated and the
condition terminate fatally. In this third group the infection is severe and persistent with increasing evidence of cardiac damage. Many of the fatalities of the acute case occurs in this division.

FOURTH GROUP - There is yet a fourth clinical type. This group consists of an overwhelming infection in which the duration of life is measured in weeks. Unless such signs as acute polyarthritis appear it is easy to error and not realize his true condition. Because of the rapidity of the course the usual signs of congestive failure do not appear and involvement of the valves may not be apparent, tho there may be a systolic apical murmur as heard in many infections but not at all characteristic. The heart often shows no signs of enlargement. The picture is that of an acute toxemia, the nature of which may not be recognized until autopsy. This is similar to the group by Davis and Weiss (18) in which death occurred with in two to 42 days.

FIFTH GROUP - Although not usually considered, one might include a fifth group. These cases would represent those patients who later in life develop signs, and at autopsy give confirmation of the presence of chronic rheumatic heart disease. In many of these, the signs and symptoms are so slight in childhood that they pass unnoticed or in some cases the memory does not recall the early childhood episodes.

Even with the difficulty involved in the division of these acute cases into clinical groups, the preceding description is very similar to the divisions given by other authoratives. (41) (31) (65) Davis and Weiss (18) present two groups that differ enough from the previous description to warrant further notice. He presents an acute or subacute heart failure with recovery, in which there is a sudden almost overwhelming onset but in which recovery occurs. The recovery is either
complete with in a few weeks with freedom from all symptom, or partial with persistant dyspnea on exertion and a limited cardiac reserve. Prognosis is difficult in this group because death may occur at anytime within 3 months to 10 years following one or more attacks of failure.

The other group consists of those patients in which the onset was gradual but in which recovery occurred even tho it was of short duration. There was relative comfort for a period of about 3 months and then failure occurred with subsequent death.

By means of this discussion of the clinical types of the acute phase, I hope to have emphasized the great variation in the course of this disease. And, in a general way to help classify any individual case. But one must keep in mind that any particular case may not adhere to any one clinical group but may present characteristics of other groups.

RECURRENTS

The great tendency to recur is a characteristic feature of the course of rheumatic infection. Because of the importance of recurrences in the course of the disease, it is only proper that they should be considered at this point. As would be expected, the incidence of recurrences is high. Lingg and Croxford (81) reported 73% in a series of 413 patients, which he adds is a minimal figure. Roth et all (54) in a series consisting of patients that exhibited carditis in their first manifestation, found an incidence of 60% recurrences.

The expectancy as to relapses has been estimated by Wilson et all (81) as follows. Up to 7 years the chances are 3 to 5 or more that a relapse will occur in the following year. From 7 to 11 the chances are about even and after 12 the chances are 2 to 5 or less. They also
found that 62% had one relapse, 19% had as many as two and 19% had three or more.

It is generally the rule that the recurrence will be of the same type of manifestation as the primary attack. However, any of the other manifestations may make their appearance. The occurrence of chorea in relapses has the same significance of mild infection that it has in the initial bout of rheumatic fever.

RELATION TO THE AGE OF ONSET - The age of the patient at the time of the initial appearance of the rheumatic infection seems to play a role in determining the frequency of subsequent episodes. In an adult group, 58.2% had at least one subsequent attack while in a group of children 78.2% had recurrences. The greatest incidence of recurrences occurred in those cases who had the first attack between the ages of 5 to 10. In this group 93.4% had recurrences. The curve after this age follows a steady downward course, but it is not until after 30 that the incidence rate falls below 50%.

<table>
<thead>
<tr>
<th>Age groups</th>
<th>Incidence</th>
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<tbody>
<tr>
<td>0-5</td>
<td>45.4%</td>
</tr>
<tr>
<td>5-10</td>
<td>93.4%</td>
</tr>
<tr>
<td>10-15</td>
<td>74.6%</td>
</tr>
<tr>
<td>15-20</td>
<td>78.1%</td>
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Of those who had two or more attacks the age of initial attack fell within the first 25 years of life, on an average. (44) It is also interesting to watch the progressive diminution in the number of recurrent attacks per individual in advancing decades. (78)

INTERVAL BETWEEN ONSET AND RECURRENCE - Lingg and Croxford (81) report that one half had relapses within one year, 66% within two years, 80% within three years and in only 15% did at least four years elapse before
the first recurrence. Mackie's figures (44) show considerable variation, 23.6% within one year, 38.8% within two years, 52% within three years and 23.8% within four years. As many as 43% waited four years or longer. The difference in the age groups studied may account for this variation in figures. In some cases there are free intervals of from 10 to 30 years before a relapse. One questions whether these are true recurrences or what might be termed reinfection. No qualified answer can be given, but they appear to be true recurrences as far as can be determined. The strongest evidence is in pathological findings. In general it is not infrequent to experience as many as four years of freedom from infection before another attack. However, it is only after a lapse of three years of complete freedom from symptoms that an individual has a better than an even chance of escaping a further bout of rheumatic fever. But even after longer periods relapses are frequent. (44)

PROGNOSIS - The significance of recurrences lies in the fact that the heart receives further strain and damage with each relapse. Even if the heart should be fortunate to escape in the first attack, it seldom is so fortunate in the second or third bout. However, the heart is usually involved in the first attack. It is generally agreed that heart disease is more common in children who have had repeated attacks. The majority of deaths from rheumatic heart disease are the direct results of rheumatic recurrences. A large number recover from relapses but there is practically always additional damage done to the heart and a large number ultimately succumb to fresh rheumatic attacks after a short lease on life. The mortality is high up to the age of 14 when it falls abruptly as does the incidence of recurrences. (57) The future of the rheumatic child depends upon his ability to weather the storm in early years.
and to reach calmer waters of adolescence when relapses are less frequent, and the power of resistance increases.

**FATAL COURSE**

The picture of fatal rheumatic heart disease in the acute phase differs profoundly from some of the early manifestations. It is frequently confused with the clinical picture of pneumonia, nephritis and other conditions. The majority are represented as an exacerbation of a long-standing clinically recognizable rheumatic infection. The duration usually extends over a period of months before the patient eventually succumbed to an exacerbation of toxic symptoms with an increase in the signs of cardiac failure. Less often the initial attack of rheumatic infection progressed to a fatal termination without a recognizable interval of significant improvement. In a few instances a fulminating recrudescence of an apparently quiescent infection ended fatally in as short a period as 10 days. In a child the course is not typical of cardiac failure, cyanosis is not seen, there is no pronounced edema or enlarged liver. The child is pale, ashen with mild edema. Death is often sudden from a terminal syncope and the general appearance is of a severe toxemia with oxygen want.

**PROGNOSIS OF ACUTE PHASE**

There are two different aspects of the prognosis of the acute or active phase of the rheumatic heart. First, must be considered the immediate prognosis of the first attack of rheumatic carditis. A few patients will succumb but the majority will survive and their future course must be investigated. The study of the prognosis divides itself into the immediate and the ultimate. On the whole, there has not been
adequate recognition of the distinction between immediate and ultimate prognosis in rheumatic heart disease. One must recognize the dangers of the acute process which may seriously jeopardize life or the patients' future activity. But one must also realize that there can be almost complete subsidence or regression of signs of cardiac damage. The heart has been set too much on a pedestal as the essential organ, while it is only one of many, and at that one of the toughest, most resistant and most capable of recovery after serious disease. Recovery is now being recognized in many cases. (76)

IMMEDIATE PROGNOSIS - It seems to be a universal opinion that in the first attack of rheumatic carditis, even though it is severe, the majority survive. Only rarely does a patient succumb to the first attack. (6) (82) However, from a study of autopsy records of 73 cases, it was found that 40% succumbed to the first attack. This would seem to indicate a much higher fatality than usually recognized. But this must be regarded with caution since it is a study of fatal cases and therefore does not include the patients that survived the first attack. Moreover, the preponderance of evidence points to the fact that the first attack carries with it a low mortality rate. The unfortunate part of this is that the majority who recover are left with a heart partly crippled (28) Those cases in which the heart is going to recover completely shows signs of such recovery within 12 months of the acute attack even tho the process may not be completed until some years later. (33) The fatal cases are rare and mostly met with in young persons and those with a strong hereditary tendency. These may be either insidious or fulminating but in either event they are usually in fragile children of feeble constitution. (51)
ULTIMATE PROGNOSIS - The future of the patients that survive the first attack of rheumatic carditis is quite varied, as has already been indicated in the review of the course these individuals may follow. Because of the extreme variability it is practically impossible to predict the ultimate course of any one individual case. All that can be hoped to be done is to acquire a thorough knowledge of the possibilities and by a careful study of each case and its progress, arrive at an estimation of the future for that case. Besides a complete study of the disease possibilities this involves experience and judgement on the part of the practitioner.

Unfortunately these individuals are all faced with the possibility of recurrences of the disease. I have already considered the influence of such attacks and some of the factors concerned with them. So their serious nature is already apparent. It might be stated, that the first attack forges a link in a chain of events that results in progressively increasing disability of the heart that sooner or later leads to the death of the patient, though there are exceptions. In some cases of slight damage the disadvantage is compensated for by the heart and such individuals may live to old age without any disability and even their normal span of life though these eventualities are distinct exceptions.

(6) Usually the child who survives the primary manifestation - I speak of a child only because of the higher incidence at this age - is able to lead almost normal lives in a relatively good state of health, attending school with average regularity and entering steady and useful employment when they grow up. However, after a variable period of months to years he has a renewed attack which leaves the heart still more damaged than it was before. It is usually at this stage that symptoms of dyspnea and palpation appear giving evidence that the heart no longer has normal
range of response. With proper treatment function may be restored but with repeated insults sooner or later a break down occurs. Each time it is more difficult to regain ground that has been lost and eventually organic and functional disturbances become so great that the patient becomes an invalid, usually years having elapsed since the first attack.

(6) The type of recurrence will help somewhat in arriving at a prognosis. Where carditis was followed by other types of rheumatic manifestations the death rate was 42.8%. In contrast to 66.6% where the recurrences were of the carditic type. (9)

So imperfect is our control of rheumatic state that we are impelled to qualify our views on the prognosis in any particular case by warning that a child is never safe from a relapse and that another attack may entirely alter our opinion. The younger the child the greater will be the possibility of such a change in prognosis will be necessary.

The early years following the initial attack clearly represent a critical period which determines in a large measure the future course of the disease. Therefore, the rapidity with which heart enlargement developed and to a greater extent the residual enlargement, serves as a reliable index of the original susceptibility of the patient's cardiac muscle to previous infection. It indicates further the muscles vulnerability to later recrudescences. It is also obvious that in these younger patients the dominant role of acute rheumatic infection as the determining factor in the cause of heart failure forces all other considerations well into the background. (5)

To return to the consideration of the early years following the initial attack. Bland and Jones (5) studied the course of events of
250 cases who died from rheumatic heart disease. In this series he found almost one half of the patients succumbed during the first 3 years after the onset. And two thirds of the fatalities occurred during the first 5 years. These figures assume increasing significance when it is realized that they have followed the living counterpart of this group of diseased patients well into the second decade of the disease.

Coombs (14) in a series of 218 patients who presented conclusive evidence of carditis found that 13 or 5.1% died within the first year. Of 204 cases traced for 5 years 25 had died, giving a fatality of 11.2% and of 117 patients traced 10 years 42 or 21.4% had died.

It would seem, that the first year is particularly fatal period following the onset. It should also be noticed that there is a fairly steady death rate year by year following the initial attack. It is interesting to note that he has followed 109 suspicious patients. In this group the
death rate was considerably different. None died the first year and
by the end of the 10th year only 6.5% had succumbed. Evidently many
of the suspicious cases did not have true carditis. David and Weiss
(18) in a study of a much smaller group of 83 cases, found that 6 died
from the initial attack, 8 more within 2 years and a total of 25 by 5
years, the remainder lived from 6 to 40 years.

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<tr>
<td>0-5</td>
<td>25</td>
<td>died</td>
<td>30% cumulative 30%</td>
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<tr>
<td>5-10</td>
<td>10</td>
<td>12%</td>
<td>42%</td>
</tr>
<tr>
<td>10-15</td>
<td>10</td>
<td>12%</td>
<td>54%</td>
</tr>
<tr>
<td>15-20</td>
<td>13</td>
<td>15%</td>
<td>69%</td>
</tr>
<tr>
<td>20-25</td>
<td>10</td>
<td>12%</td>
<td>81%</td>
</tr>
<tr>
<td>25-30</td>
<td>5</td>
<td>6%</td>
<td>87%</td>
</tr>
<tr>
<td>30-35</td>
<td>7</td>
<td>8%</td>
<td>95%</td>
</tr>
<tr>
<td>35-40</td>
<td>3</td>
<td>5%</td>
<td>100%</td>
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Ash (2) observed the course in 445 children. His figures are consid-
erably higher for this early period. Of those who died, 33% died within
the first year, 51% within 2 years and 74% within 5 years of the onset.
This consisted of the deaths that occurred over an average of 7½ years.
The first 5 year period is also important in that beyond this period
there is less likelihood of reactivation and hence progression of cardiac
disease.

PROGNOSIS RELATED TO DEGREE OF INVOLVEMENT - The patient with definite
early signs are not much more likely to become permanent cripples than
those with doubtful signs, but they are more likely to die off at an
erlier age. (14) Wilson (47) found that a child who does not present
any recognizable symptoms of active carditis may be expected to show a
lesser degree of cardiac involvement in later life and to have a longer
life. Virulent attacks of carditis, experience proves, do irrepair-
able damage and recurring attacks of less severity are much more grave
The degree of impairment depends upon the severity of the damage to the valves and the myocardium. A slight mitral insufficiency may be borne without any evident disability. Such a patient may go thru life with no symptoms of failing cardiac circulation. Ordinarily the damage is severe enough to put permanent handicap upon the efficiency of the individual. Thus, in patients presenting these symptoms, 55% died within one year after the onset, and as high as 27% showed cardiac symptoms and signs of less than one month duration. In well definable cases a certain proportion die from heavy infection with myocarditis while survivors develop valvular disease with some permanent myocardial injury. In the suspicious group there is infection with transient myocarditis and gradual development of true valvular lesions.

**PROGNOSIS - WITH THE DEVELOPMENT OF HEART FAILURE** - The fact that in many cases the duration of life is short following the appearance of congestive failure is significant. In some cases the exact time of onset was not clear either because it developed insidiously or because a neurosis had developed in childhood from the knowledge of heart disease. In a series of about 130 patients, 22 died within 2 weeks from the onset with definite cardiac symptoms, 36 -37% - died within one month, 62 within 6 months, 78 -56% - within one year, 44 lived 3 years or more, 19 for 5 years and 5 cases for 10 or more years. When a patient gives a history of frequent periods of congestive failure with evidence of continual activity there is no real hope. Further, if after careful treatment for months, evidence of failure persists the outlook is bad even though the rhythm remains regular. (29) After the onset of congestive failure the prognosis is grave. Less than one half will live out the
following year.

VARICUS FACTORS IN PROGNOSIS - In only a limited extent, are physical findings of value in prognosis of life or in determining the degree of disability which will remain after the attack subsides. (71) It is important to realize that children and young adults develop cardiac failure because of an active rheumatic process, mechanical failure from a previous rheumatic episode virtually never occurs. (72) In 94%, representing most unfavorable type of reaction, extensive cardiac enlargement occurred early and was noted at the time of the initial attack in all of those who died within one year of the disease. (5) Some patients die without enlargement of the heart while others live with enlarged hearts in all comfort. Yet as a general rule, those with enlarged hearts are under a distinct handicap as far as his chances of recovery from active infection are concerned. And even if he succeeds, his cardiac reserve is lowered. (71) An infection, so slight as to pass unnoticed in a normal sized heart might be sufficient to precipitate heart failure in a markedly enlarged heart. The prognosis thus depends upon the size of the heart to a certain extent. (72)

From clinical experience observers have gathered the impression that the age of puberty represents a favorable landmark in the natural course of the disease. (5)

<table>
<thead>
<tr>
<th>Age onset</th>
<th>1-5</th>
<th>6-10</th>
<th>11-15</th>
<th>16---</th>
</tr>
</thead>
<tbody>
<tr>
<td>patients</td>
<td>60</td>
<td>132</td>
<td>36</td>
<td>16</td>
</tr>
<tr>
<td>duration</td>
<td>4.9</td>
<td>5.0</td>
<td>5.0</td>
<td>8.0</td>
</tr>
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</table>

These figures show an increase in the average years of duration in the group whose onset was beyond puberty. There are too few patients in this group, however, to warrant any definite conclusion but this agrees
with the clinical impression of others. (82) Swift (68) states that a condition of resistance seems to develop at about this time. Lingg and Croxford (81) also agree that after the age of 12 it seems that an immunity develops. However, he also points out that the ages of 10 to 14 appears to be a critical period as judged by mortality and increased activity. Schlesinger (66) stresses his conviction that puberty is the determining period at which the power of resistance to rheumatic infection increases, cutting down on the number of recurrences.

The type of environment seems to play a small part in the prognosis. The general experience is that the better the child is cared for in regards to food, clothing, exposure and fatigue the greater the chance of warding off recurrences. (9) (58) However often the most severe case is in a child growing up in the ideal surroundings. One can hardly doubt that the decrease in poverty due to the betterment of living conditions under which industrial population has to live has been to some extent responsible for the decrease in fulminating cases. Although other factors play a large part. (14) (9)

There is quite good evidence that people who have something to do make a better fight than those without any obligations. This is more true for private patients, probably because the "better-to-do" class can rest at intervals when they feel like it while the working class has to keep going. There is a stage during adolescence when the character of the work done by the patient has a vital import. During adolescence many who have had bouts of rheumatic infection are cast upon the world without either physical or educational resources to enable them to keep pace with their healthy contemporaries. At the same time they must fight their infection along with shortage of food, long hours and exposure. Their lot is anything but hopeful. (14)
There is a definite belief now that hereditary tendencies play an important part in the incidence of rheumatic infection. (26) However, the question arises as to what effect the hereditary tendency has to do with the prognosis and severity of the infection. Nothing definite can be said, but the type and the degree of the disease appears to be more severe than in the general run of cases. (14)

Race also seems to have some influence. From reports there appears to be an apparent resistance among negroes to rheumatic infection. Ash (2) found that in the clinic of the Children's Hospital Philadelphia where the white and negro children are about evenly divided the rheumatic children were about 83% white and only 17% negro. Wilson (31) and her co-workers found that only 4.5% of the children were colored although the clinic draws from a colored district. Sutton shows that there seems to be a susceptibility to rheumatic heart disease among the Italians while the Jews are afflicted to a degree less than the general clinic percentage. The Irish apparently have a rather high percentage. (66)

As regards sex the girls were more frequently and more seriously affected than boys. (9) Also the children living in changeable climate of the temperate climates were more seriously infected than those who live in the more temperate zones. This last factor is probably due to the greater incidence of recurrences found in climates with changeable weather. (9)

FUNCTIONAL LIMITATION - It is impossible to express in precise terms an outline of the influence of the rheumatic heart disease on functional activity of the patient. However, certain generalizations have been made by Coombs. (14) First, those who died during the early course of the disease were quite severely handicapped during the period
immediately before death. This was partly due to recurrences which cut them off from school and normal activity. By the time these patients get out of school they usually have signs of advanced cardiac damage and decompensation which greatly limits their activity. It is a reasonable generality to make; that no one whose heart is extensively damaged by the time of puberty is going to be fit for work, except of the lightest form and that not for long. On the other hand, there are those who recover and live a normal life. Just short of complete recovery, is the career of the patient who has no symptoms. He competes without being aware that he is handicapped and indeed sometimes without breaking down under the strain. Often it is only by chance that the chronic form of the lesion, to be discussed later, is found at some examination. Of recoveries with limited measures of efficiency, there are every degree. These limitations, however, usually begin to appear somewhere before the age of 30. There are extraordinary degrees of variation not dependent solely upon the nature and extent of the lesions with their encouchment on functional efficiency. Incalculable elements of courage comes into play and upsets all accuracy of prognosis. Some individuals can stand much more than others. Just as in the case, where because of an injury to a leg it must be amputated, one individual will get a wooden leg and go about his business another will get a tin cup and sit on a corner asking for alms. In some cases the kind of work with its requirements on the individual will determine to what extent he will be limited.

SUMMARY - One must remember in the prognosis of each individual case that the majority do recover and that many will do well for years to come, under good sensible care. Even heart murmurs both systolic and diastolic
and heart enlargement may disappear with the subsidence of cardiac dilatation due to rheumatic myocarditis. The presence of pericardial involvement is often of relative unimportance though it does indicate a severe infection. Cutaneous nodules are usually of more significance. Of course, one must recognize the ultimate prognosis is in many of these cases not good. Only time will tell when another acute rheumatic infection may add its toll of strain to the already damaged heart. During the acute rheumatic carditis one should keep an open mind about both the immediate and ultimate prognosis no matter how sick the patient may be at that moment. The functional state of the circulation and the cardiac reserve are the important factors in the prognosis of persons with cardiac damage. (76)

Coombs (14) has presented the theoretical course based on his investigation of 300 cases seen in the early stage of this disease. In one third of these cases the diagnosis must be considered as suspicious. Since his discussion presents a very good summary, I would like to present it at this point in the discussion. Out of 300 patients, 50 will fail to survive the first 30 years. Of the survivors, 115 will have lost all physical signs, 75 will show a true recovery and 40 were probably diagnosed on inadequate grounds. The remaining 135 will reach maturity with cardiac lesions becoming quite definitely evident between 20 and 30 years of age. Some limitation of power and work will have appeared in the form of dyspnea, at first with only exertion, but later evident at all times. By 30 years there will be only 114 survivors, by 40 only 82 and by 50 the survivors number only 60 all of whom will have a condition of invalidism so complete that only one fourth will be capable of work.
I would like to leave the concept that although there are general rules which are helpful in the prognosis of heart disease, there is nothing, perhaps, even now more difficult in practice of medicine than the prognosis of cardiac disease. A distinct advance can be affected by recognizing that immediately serious and grave cardiac states may be followed by functional recovery so complete that ultimate prognosis is good for many years to come. (76)

POTENTIAL HEART DISEASE

Up to this point we have found that a few have succumbed to the early years of the disease, but the majority have survived. Of these that survive a few will lose all physical signs of heart disease, while the majority will reach maturity with cardiac lesions. At this stage, I would like to consider the prognosis of the young individuals between the ages of 12 and 30 years, who have had rheumatic fever and and present on physical examination either an entirely normal heart or at the most an apical murmur. This group has been far less thoroughly explored than the preceding and the subsequent groups. And few indications have been set up showing the future course of these patients with slight valvular damage. Numerous discrepancies have appeared, resulting from two difficulties. First the faulty memory of older patients with respect to their early rheumatic histories, second the difficulty of trying to follow closely a large group of essentially well people over a long period of time, have added to the trouble in studying this group.

DEFINITION - Boone and Levine (7) have divided this group into two different divisions, because of the different findings and the subsequent
developments. The first is that of potential heart disease which is diagnosed on the basis of a normal heart or one with a grade one systolic murmur following a history of rheumatic infection. The second group consists of those with a history of rheumatic infection and a systolic murmur of grade 2 or better. This latter group they labeled rheumatic mitral insufficiency. While both of these can be considered as potential heart disease, I would like to divide the group into these two divisions.

POTENTIAL HEART - Patients in this category live a normal life without any symptoms of cardiac disease. They may be even entirely unaware of the underlying condition. It is important to know the future course these patients may travel. In a series of 166 cases of potential heart only 8 or 4.8% subsequently developed recognizable valve lesions or a serious question of it. It is interesting that there seemed to be little difference in this respect between single and repeated attacks of rheumatic manifestations. The group having repeated attacks showed only a slightly higher incidence of valve deformity. There is a 94% chance of escaping valvular disease if less than 10 years have elapsed since the first attack and practically 100% after the passing of 10 years. Following a single attack of rheumatic infection, those patients who could be diagnosed as potential heart disease have about a 96% chance of escape and if after 5 years following the attack nothing new develops there is practically a 100% chance of them escaping serious damage. (7)

Lingg and Croxford (81) found in a series of 159 children classified as potential heart disease, 37.1% of the cases developed definite heart lesions. The remaining 61.6% stayed in the potential heart disease group. St Lawrence (63) in a series of 65 cases observed for an
average of 41 years, found that 75% remained free of cardiac disease and 25% contracted a cardiac lesion. In both of these last series there is a higher percentage than given by Boone and Levine. But it must be remembered, that in comparing figures from different series of cases, variation will occur due to the method of examination, the criteria used and the age group studied.

In tracing what changes may take place during the following years, we find that the patient may remain well and may never have a return of rheumatic infection though he may show a slight systolic murmur on examination. Occasionally this murmur may gradually diminish in intensity. Rarely, it may disappear entirely. He may, therefore, live his normal life and never be embarrassed by his heart. This is perhaps the exception unless with recovery no murmurs whatever remain. On the other hand, this patient may either as a result of recurrent bouts of rheumatism and reinfection of the heart or possibly because of the inherent nature of the original infection with subsequent chronic progression, develop signs of mitral stenosis or other chronic valvular lesion. In a few cases, accidents such as subacute bacterial endocarditis or pneumonia will occur and be the cause of the patient's death. (39) It is difficult to escape the conclusion that these cases of potential heart disease represent a group whose hearts were relative resistant to rheumatic infection. (7)

MitraL INSUFFICIENCY - It is maintained by certain authors that organic mitral insufficiency does not exist, or it is extremely rare. It is, of course, recognized that there are many instances of benign systolic murmurs and that the dire prognosis given in former years was responsible for a great deal of harm. But it is wrong to deny the existence
of such a non-fatal condition by the use of autopsy data. (39) This is usually the earliest sign of permanent damage to the heart by the rheumatic state. It may even be apparent a few weeks after the infective period. However, the heart soon accommodates and the patient may carry on for years without any symptoms of cardiac damage. (6)

In a series of Boone and Levine (7) 58% of the cases persisted virtually unchanged throughout the period of observation, while 42% subsequently developed serious valvular disease. In those cases following a single attack of rheumatic infection there was found to be a 81% chance of escaping further lesions during the first five years and almost 100% after 5 years. In patients with a history of repeated attacks there was only a 39% chance of escape under 5 years and a progressively better prognosis the further removed from the initial attack, during the next 20 years.

Unfortunately because of the great susceptibility to reinfection these cases rarely remain stationary and in the course of time mitral stenosis develops or even multi-valvular lesions appear. (62) As long as it is only a regurgitation, the patient is apt to be in good health. Even this, however, is not invariably so, for some cases die from heart failure showing a markedly dilated heart and mitral insufficiency without stenosis. On the other hand, the condition usually develops into mitral stenosis which eventually is fatal, although for many years there is only an incompetency. (62)

The conclusion is that although the diagnosis of organic mitral insufficiency should be made with caution, it is a condition that actually exists, especially in young rheumatic individuals. And that this condition generally develops into mitral stenosis after a period
of time, although some cases will recover entirely and live a normal life.

CHRONIC VALVULAR HEART DISEASE

Now is the time to discuss those cases which may be regarded as the crippled survivors of the previous groups. As in the preceding divisions, there is a great variability in the course these patients will run. One patient with valvular heart disease carries on in comparatively good health for many years, and another who starts with seemingly same lesion succumbs in a short period of time. Death is usually ascribed just to heart failure while a closer analysis of recent events often reveals that there is a more specific complication which cannot be regarded as ordinary circulatory failure. In other words, all patients with rheumatic valvular disease do not die in the same way. (35) The causes of death will be considered more in detail later.

In chronic valvular disease the primary valve involved is the mitral. If other valves are involved they are usually in association with this one, though occasionally one of the other valves is affected alone, in which case it is usually the aortic. When the mitral valve is involved along with other valves, the mitral lesion is the greatest in extent. (3) The incidence of mitral valve involvement and the combination of mitral and aortic is given in the table below. (19)

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<tr>
<td>mitral</td>
<td>62.5</td>
<td>51.5</td>
<td>77.0</td>
<td>44.0</td>
</tr>
<tr>
<td>mitral and aortic</td>
<td>28.6</td>
<td>19.2</td>
<td>9.3</td>
<td>39.2</td>
</tr>
</tbody>
</table>

Mitral or aortic with pulmonary or tricuspid, occurred in only 4.5% of cases. (19) The aortic valve was involved alone in about 4.1%. (6)
Thus the mitral valve is by far the most important.

The age of onset has very little effect upon which valve will be affected, though there is a slight tendency for aortic, tricuspid or pulmonary lesions to occur more often in cases which have the initial attack early in life. (19) Multiple valvular lesions were greatest in children who had numerous attacks of carditis while the mitral valve occurred alone more often with one or only a few attacks of carditis. (81) In a history of chorea alone, no cases of valvular involvement were found. Where there was history of carditis and chorea there was no difference in the valvular lesions. (7)

DEVELOPMENT OF MITRAL STENOSIS - The development of chronic valvular disease can best be followed by tracing the development of mitral lesions. Some cases develop mitral stenosis without any previous history of rheumatic infection. However, these individuals usually have forgotten early attacks, or the early attacks were very mild and passed unnoticed. The majority, recovered from the attack of acute rheumatic fever with no evidence of cardiac damage, or at the most a systolic murmur. After a prolonged period of apparent freedom from activity, they develop mitral lesions. Because of the frequency with which the morbid process in the heart progresses in the absence of any criteria of acute disease, they require guarded prognosis and prolonged, careful observation. (44) The first indication that the mitral valve is becoming stenosed, is a snapping quality, or accentuation of the first heart sound at the apex. Later the characteristic murmur found in mitral stenosis gradually appears. (39)

How soon these changes occur is a variable matter. Upon rare occasions the signs of mitral stenosis developed within one year,
though generally many years elapse between the original infection and the appearance of definite evidence of mitral stenosis. In a series of cases, Mackie(44) found 86% developed physical signs of stenosis after two years from the acute attack. The longest period in his series was 6 years. In another series, one third of the patients developed valvular disease in the course of 10 to 15 years after the onset. Again arises the difficulty in comparing figures in different series of cases. However, as a rule many years elapse before signs of the lesion are detected. Even though mitral stenosis usually appears in young adults, a few cases are seen after 50 years of age.

COURSE - It isn't the purpose of this paper to discuss such an entity as mitral stenosis in complete detail. But in so far as this is an important part of the rheumatic heart a short summary will be given. The great majority of persons who have had rheumatic fever - about 85% - develop chronic deforming valvular disease. This generally begins in childhood, though it is more commonly found in the decade from 15 to 25 years and is relatively infrequent after 50 years of age. (11) The ultimate forecast may be said to be almost always unfavorable. It handicaps activity and is often fatal before or after the patient reaches adult life. The strain on the heart is apparently very great, no matter how good the condition of the muscle may be. In older persons with long standing valvular defects, heart failure is most often not due to exacerbation of the infective process. However, we must remember that initial attacks may occur in late years, even after 60. The cause of heart failure is essentially the same as in hypertension. There is an inadequacy of metabolic exchange of the hypertrophied muscle. Also, the residual damage from myocarditis that was present years earlier is
a factor. It must be remembered that the functional capacity of the heart is decreasing from the changes incidental to advancing years. (23) Auricular fibrillation and congestive failure eventually lead to death. But now and then bacterial endocarditis, or an intercurrent infection is the cause of death. (11)

PROGNOSTS - As has already been indicated, the diagnosis of chronic valvular heart disease is a serious situation. The question arises as to what importance the type of valvular lesion plays. There actually seems to be very little relation. Though, some believe that the cases of aortic stenosis are longer lived and that the pulmonary and tricuspid lesions render the prognosis less favorable. (19) The average term of life after the onset of the initial infection was about the same regardless of the type or number of valves involved, according to David and Weiss. (18)

Cabot (10), however, presents figures that show a definite relation between the prognosis and the type of valvular lesions. In thirty six cases of pure mitral stenosis, there were 27 that lived as long as 10 years or more. Moreover, 6 lived 25 to 35 years following the onset of the disease. The average duration in this group was 15 years. In these mitral cases 30% survived 50 years of age, though the danger point in from 30 to 40. On the other hand, the term of life in combined mitral and aortic lesions, is strikingly shortened. In more than one half of his cases the disease killed within one year with an average duration of 3 years against 15 years for patients with pure mitral defect. Only one case lived more than 10 years. In other combined lesions the prognosis seems to be better than in the preceding case, but it is still worse than the pure mitral cases. Nine out of 28 lived 10 years or more.
Only two died within the first year, the average in this group was 10 years. The patients with aortic stenosis proved to be the longest lived group. All of 76% lived beyond 40 and 50% survived the 50th year.

According to Coombs,(14) up to the age of 25 aortic involvement was deadlier with 16.1% of deaths against 12.4% of mitral deaths. At 30 years, they were neck and neck and at 40 the percentage of mitral deaths was 60.9 while aortic was 52.2%. The early appearance of aortic lesions indicates a more severe and extensive infection, thus accounting for the higher incidence of deaths. In later years, the heart with pre-dominately aortic signs, runs a relative benign course.

With the appearance of mitral stenosis in the young patient the prognosis becomes ominous. The outlook is bad and the death rate is almost 35%. (2) (57)

In general, as regards prognosis its essential idea is that the subsequent course of each patient is to be foretold not so much by the presence or absence of signs of valvular disease or by the type of valvular disease thought to be present. But rather by the degree of cardiac enlargement and the grade of cardiac failure. (19) Austin Flint (24) writes that the significance of murmurs is limited to the existence of lesions and their location. They give no information as to the prognosis. The size of the heart is of greater importance. There is a vast amount of positive harm being continually done to the patient by taking too seriously the prognostic significance of the valvular lesions. Prognosis in heart disease still lacks definition and remains largely a matter of individual experience and opinion.

In histories of 1000 men followed for 10 years, one of the most striking features is the large proportion of the cases, especially those
with uncomplicated valve defects, who live through the 10 years uneventfully and without change in the physical signs. Moreover, even those who die within this period, the majority showed no steady progress to a fatal end. The original condition remained stationary year after year until death occurred suddenly or until some intercurrent infection, auricular fibrillation or unknown cause led to congestive failure and death within a relative short time. Finally, there are exceptional cases in which definite rheumatic valve lesions are present for as long as 30 years or more without heart failure developing. These cases usually succumb to an independent ailment. (23)

In the group with little or no enlargement and good or fair exercise tolerance the prognosis is good. Only 21% died while 52% survived uneventfully without change. Then there was poor exercise tolerance with moderate enlargement, the outlook is poor. Though a small proportion remain unchanged, about one half died in the 10 years of observation. When there is great cardiac enlargement or signs of congestive failure, prognosis is very bad. About 80% died within an average duration of two and one half years. (27)

MORTALITY AND MORBIDITY

Unfortunately, the extensive studies made on mortality have not been divided into the groupings with which I have chosen to discuss the rheumatic heart. However, many have been divided into young and old age groups. Roughly these correspond to the acute and chronic divisions which I have used. Because of the lack of complete correlation, I have decided to devote a separate section on this feature of the discussion.
atic condition, the development of cardiac symptoms occurs with considerable variation. The date at which a patient begins to suffer from symptoms due to impairment of his heart is an important landmark. Increased fatigue and dyspnea constitute the chief evidence. (39) The clinical phenomena are such as to change a patient from class I to class II a or b. Symptoms of cardiac insufficiency are experienced by nearly all patients. In 96.2% the mean interval between the onset of infection and the first clinical evidence of cardiac insufficiency was 11 years. It is noteworthy that 22.9% of patients presented symptoms within a year after the first manifestation. In more than 50% the free interval was less than 10 years. The older the patient when the first infection with rheumatic fever the shorter the interval before cardiac symptoms made their appearance. (20)

Gerheardt, studied 122 cases for the interval of time between the initial onset and the appearance of heart symptoms. He found an average period of 12 years in this series. However, this does not fully represent
the situation because there is a wide variation in the number of years elapsing.

AGE SYMPTOMS APPEARED - Other investigators have studied the time of onset of cardiac symptoms in relation to the age of the individual at the time they appear. Simmons (60) with 155 cases found a progressive increase in the number showing symptoms with each decade up to the age of 40. The greatest number were between the 2nd and 5th decade, amounting to 58% of the cases. Six did not develop symptoms until after the 50th year.

<table>
<thead>
<tr>
<th>Age of patient</th>
<th>1-10</th>
<th>10-20</th>
<th>20-30</th>
<th>30-40</th>
<th>40-50</th>
<th>50-59</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-10</td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>20-30</td>
<td>26</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30-40</td>
<td>32</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>40-50</td>
<td>10</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>50-59</td>
<td>6</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Cabot (10) who started with post mortem lesions and worked back to the clinical findings, found that in 67% out of 63 cases heart disease began before 30th year. When compared to ambulatory patients on basis of clinical history it was found to be 82%.

ONSET SYMPTOMS TO ONSET HEART FAILURE - Heart failure has been taken to exist when the symptoms of cardiac insufficiency are so pronounced that the patient is unable to do any work, but has to go to bed and remain there for some time. In general patients began to experience attacks of heart failure about two years after the symptoms of cardiac insufficiency had appeared. However, this group included such accidents as bacterial endocarditis and intercurrent infection so is definitely shorter than it would otherwise be. (20)
Interval between onset symptoms and first attack heart failure

DURATION AFTER ONSET HEART FAILURE - Fishberg (23) gives us an idea on the duration of life after the onset of heart failure. After the symptoms of heart failure set in, the course is very variable. Some get on well for even 5 to 10 years. Others have repeated bouts of heart failure during a period of years, from each of which they recovered sufficiently to be up and about. However, the tendency is toward longer duration and less satisfactory improvement after each successive attack. In other words each successive attack is more severe than the preceding ones. Such a protracted course is more common in those who lead a comparatively leisurely life than in the hard working patient. The prognosis is poor in rheumatic heart disease once it has resulted in congestive failure enough to require bed rest. The average duration after onset necessitating bed rest was found to be 2½ years. DeGraff and Lingg (20) found the mean duration of life was 3 years. In 75% less than 5 years. On this basis, the chance that a patient will be alive one year after the onset are even, that he will be alive 5 years are 1 in four.
More than 50% of Davis and Weiss (18) patients succumbed within one year after the onset of definite heart failure; (total of 78 patients) 44 patients lived 3 years or more; 19 for 5 years and 5 cases for 10 or more years. This later series consisted of younger patients than the previous. The conclusion is that once a patient is in heart failure his course is about run, and at the best he will live for only a few years.

DURATION OF LIFE - This can be settled only with approximation to accuracy. We may surmise that the disease began at the date of the first, often only attack, of acute rheumatic fever or chorea. The fatalities occurring during the early years after the acute attack have already been discussed. This section will deal essentially with those individuals who live beyond this stage.

Willius (78) mentions that the duration of life is much shorter in cases of patients who receive the first infection late in life than in those whose first attack occurs early in life. The patients whose initial
attack occurred in the 1st and 2nd decades of life lived 25 to 32 years, while when the attack occurred in the 5th and 6th decades they lived only 10 to 11 years.

Aastrup (1) found the average duration of life to be 25 years. Cabot (10) found 35 or 40% of their patients lived 10 years or more. DeGriff and Lingg (20) found that the mean duration of life, from the onset of rheumatic fever to death was 15 years. Although some patients lived much longer - one as long as 58 years - such experiences are rare. Seventy five percent lived for less than 23 years after the onset. It may be said then that a patient has only on even chance of living 12 years and only one chance in 5 of living as long as 25 years after the initial infection. The mean duration in the above series is much shorter than the 30 years estimated by Coombs (14) His method of linking two groups together is questioned as to its accuracy. Willius with much the same method as used by DeGriff and Lingg arrived at a mean duration of life of 14 years, corresponding well with the latter's findings. Grant, (27) showed that one half of his group died within 10 years. But his cases were limited to men, the interval of 10 years included all stages of the disease not a progression of any one and lastly he divided them according to valvular lesions without respect for etiology.

Once the patient complained of symptoms of cardiac insufficiency, life expectancy is brief. The mean duration was 5 years, 50% died within 3 to 4 years.
AGE OF DEATH - This will vary considerably in different studies because of the variables already mentioned. DeGraff and Lingg (20) in a series of 644 patients, three fourth of whom had died at the time of the report, found that the mean death age was 33 years with the majority occurring between the ages of 23 to 42 years. Even though it is possible for a patient to live for years, even beyond 70, three fourths do not survive much beyond the age of 40 and about one half are dead at 53 years. If the patient contracts the disease in adult life or survives the first 2 decades of life death is most common between the ages of 30 and 39, at which time about 30% die. Only about 10% survive the 5th decade.

Willius (78) found that the average age at the time of death was 32. This was slightly higher when the first attack occurred in the second decade. Aastrup (1) found the average age to be 45.5 and accounts for its being higher than DeGraff and Lingg because the latter traced only 73% of the series to their death and that on the whole their patients were rather young. Simmons (60) found the average age to be 36 years. Thayer (69)
putting it another way, found that 75% occurred in the first 2 decades of life and that 88% occurred in the first three decades of life. According to Davis and Weiss (16) there are few deaths in the first decade. The deaths are distributed mainly over the second to the seventh decades. The largest number are found in the fourth and fifth decades. But as many as 23% occurred in the sixth and seventh.

If one limits himself to younger patients the age of death will be much different, because here must be included the deaths due to the acute process. In such a group, Poynton et al. (50) with analysis based on 350 fatal cases found the death rate was worse between 6 and 12 years and the majority of deaths occurred before 20 years. Bland and Jones (5) on analysis of 306 patients with the ages not exceeding 21 years and inclusive of only the first 10 years of the disease found the average to be 8 years. Wilson (82) found that 85% died before age of 17 and the average age was 12.6 years with greatest no. in the range of 12 to 16 years inclusive.

CAUSE OF DEATHS - A search for the exciting cause of heart failure showed that in the first five decades of life an active infection was
found with significant frequency. Adults with mitral stenosis even without a history of rheumatic fever showed an active myocarditis of recent origin in a high percentage of the cases, and many a well developed valvular lesion. The studies show a striking correlation between heart failure and activity. This was true even with those dying of circulatory failure as late as the fifth decade. (55) Coombs (63) has expressed the belief that the onset of failure may be traced to an active myocarditis in apparently quiescent cases. It is not sufficiently appreciated that in the cases of rheumatic fever in adults of the 3rd, 4th and even 5th decade, that a recurrence of rheumatic myocarditis rather than a healed mechanical defect may in a majority of cases be the precipitating cause of circulatory failure. In a few instances complete quiescence was present as early as the second decade of life. In individuals with valvular defects, failure was found in a majority of cases, to be precipitated by expected contributory causes occurring at this time of life such as hypertension, atherosclerosis of coronaries, coronary thrombosis, and myocardial degeneration. It is equally important to notice that a number of individuals who have had rheumatic heart disease can become completely quiescent and reach the 5th to 6th decade of life. And then die a totally unrelated death without evidence during life of myocardial failure directly attributed to mechanical defects of rheumatic origin. (55)

<table>
<thead>
<tr>
<th>age</th>
<th>active</th>
<th></th>
<th>quiescent</th>
<th></th>
<th>total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>no.</td>
<td>%</td>
<td>no.</td>
<td>%</td>
<td>no.</td>
</tr>
<tr>
<td>1-10</td>
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<td>22</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>11-20</td>
<td>41</td>
<td>95</td>
<td>38</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>21-30</td>
<td>11</td>
<td>78</td>
<td>11</td>
<td>5</td>
<td>22</td>
</tr>
<tr>
<td>31-40</td>
<td>21</td>
<td>70</td>
<td>21</td>
<td>9</td>
<td>30</td>
</tr>
<tr>
<td>41-50</td>
<td>8</td>
<td>38</td>
<td>8</td>
<td>13</td>
<td>62</td>
</tr>
<tr>
<td>51-60</td>
<td>2</td>
<td>13</td>
<td>2</td>
<td>13</td>
<td>87</td>
</tr>
<tr>
<td>61-70</td>
<td>1</td>
<td>12</td>
<td>1</td>
<td>8</td>
<td>88</td>
</tr>
<tr>
<td>71-80</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>4</td>
<td>100</td>
</tr>
</tbody>
</table>
Poynton et al. (50) have divided their series into those cases under 12 years and those over 12 years of age. The following table gives an idea as to the cause of death in these two groups.

<table>
<thead>
<tr>
<th>Under 12 years</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>22 died</td>
<td>died with acute carditis</td>
<td></td>
</tr>
<tr>
<td>11 died</td>
<td>without evidence of acute carditis</td>
<td></td>
</tr>
<tr>
<td>7 died</td>
<td>with malignant endocarditis</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Over 12 years</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>29 died</td>
<td>of mitral stenosis</td>
<td></td>
</tr>
<tr>
<td>22 died</td>
<td>of malignant endocarditis</td>
<td></td>
</tr>
<tr>
<td>20 died</td>
<td>of mitral stenosis and regurgitation</td>
<td></td>
</tr>
<tr>
<td>15 died</td>
<td>mitral and aortic</td>
<td></td>
</tr>
<tr>
<td>9 died</td>
<td>acute carditis</td>
<td></td>
</tr>
<tr>
<td>4 died</td>
<td>predominant aortic</td>
<td></td>
</tr>
<tr>
<td>1 died</td>
<td>dilatation of heart and external adhesions</td>
<td></td>
</tr>
</tbody>
</table>

These figures show preponderence of acute carditis in the young patients and of valvular lesions in the older patients. Cabot (10) gave the mode of death in a series of 146 patients.

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Passive congestion</td>
<td>74</td>
</tr>
<tr>
<td>non cardiac disease</td>
<td>40</td>
</tr>
<tr>
<td>Embolism</td>
<td>12</td>
</tr>
<tr>
<td>Acute sepsis</td>
<td>18</td>
</tr>
<tr>
<td>Sudden unexplained</td>
<td>2</td>
</tr>
</tbody>
</table>

Stroud and Twaddle (66) give the following summary of the deaths in 144 patients who were between the ages of 5 and 28.

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Congestive failure</td>
<td>104</td>
</tr>
<tr>
<td>acute rheumatic infection</td>
<td>11</td>
</tr>
<tr>
<td>Bacterial endocarditis</td>
<td>9</td>
</tr>
<tr>
<td>Sudden death</td>
<td>9</td>
</tr>
<tr>
<td>Non cardiac deaths</td>
<td>11</td>
</tr>
</tbody>
</table>

SPECIFIC CAUSES OF DEATH - From the preceding presentation one gets a fairly good idea of the types of death which occur in the patients with rheumatic hearts. So now we will take up each of these separately. As is indicated above many deaths that occur in this type of patient are entirely an accident and unrelated to the cardiac condition. For this and other obvious reasons these will not be discussed.
CONGESTIVE HEART FAILURE - About 33% or 49 cases, in a series of laws and Levine (35), died as a result of congestive failure. The largest number of deaths were in the 5th decade. The sexes were equal in number. Twenty four gave a previous history of rheumatic fever, four of chorea, four of both and 17 a negative history. Rhythm in this group was frequently irregular, reflecting the great importance of auricular fibrillation, as an underlying factor in the rheumatic patient who succumbs to congestive heart failure. This is especially true when mitral stenosis alone is present, only two dying with a regular rhythm. The duration of life, on an average, was 4.2 years after the appearance of dyspnea. Where more than one valve was involved the duration of life was somewhat greater. Thus the finding of more than one valvular involvement does not mean a poorer prognosis. Moreover, it may be expected that the length of life may be longer. But, on the other hand, although the average duration of life after dyspnea may be greater, the age at death is younger than when one valve is involved.

ACUTE RHEUMATIC CARDITIS - The second most common cause of fatalities was the acute rheumatic infection. The clinical course these patients run has been fairly characteristic. There was usually persistent moderate fever, a rapid heart rate and anemia. Over one half, developed congestive failure during the acute attack and the usual therapeutic measures were of no avail. Almost one half were under 20 years of age and two thirds under 30. This group comprises the youngest patients and it is in these that rheumatic fever as a direct cause of death is most feared. It is evident that patients with a past history of chorea alone, rarely succumbed to an acute rheumatic carditis. The predominant pathological change was an extensive damage of the myocardium. In practically every case the rhythm was regular. However, when auricular fibrillation was present, there was also a mitral lesion
As compared with the group of congestive failure, the length of life after the onset of dyspnea and other evidences of circulatory failure was on the whole rather short. This does not refer to the duration after the acute process began, as this was very short, often being only weeks. (35)

EMBOLI AND THROMBI - This group consists of those in whom an embolus or thrombus of a vessel was the determining factor in the cause of death. Embolism, is not always responsible for death. It is well appreciated that complete recovery or partial clinical recovery with life of years duration following an embolism is not a rare occurrence. Out of 164 fatal cases of rheumatic heart disease, 10% of the deaths were caused by embolism, in an additional 5% the embolic accident definitely contributed to the occurrence of death. In 6% there was suggestive, but not conclusive evidence that embolism contributed to the cause of death. In about 61% of the instances of fatal embolism the accident occurred in patients with slight or no evidence of circulatory failure up to the time of infarction. In the remaining 39% there existed a considerable degree of insufficiency. (73) In the series of Laws and Levine (35) they found a similar incidence of embolism causing death. It amounted to 11.5% against the 16% above. Following the occurrence of embolism, particularly in the brain, lungs and extremities, the existing circulatory insufficiency developed in patients with previously normal circulation. It is apparent that embolism is a so called accident in rheumatic heart disease, causing death either directly or contributing to it. The patient might have been expected to live considerably longer if this accident had not occurred.

This type of death occurred in the patients with mitral stenosis,
probably because auricular fibrillation has the highest incidence in these cases. Embolism results from the dislodgement of mural thrombi which have formed in the auricles during the course of auricular fibrillation. The rhythm in each instance except one was irregular. This is a striking confirmation of the prevailing impression that auricular fibrillation is an important predisposing factor in the formation of mural thrombi with subsequent embolism.

SUBACUTE BACTERIAL ENDOCARDITIS - There was a surprisingly large number of cases presenting this type of death, practically 29%. (35) The incidence found by David and Weiss (17) revealed that one case of subacute bacterial endocarditis occurred in every 10 cases of rheumatic heart disease or approximately one in every 4 cases of fatal rheumatic heart disease. The acute bacterial endocarditis occurred in one out of every 25 cases and one in nine of the fatal cases. The possible relation of subacute bacterial endocarditis to rheumatic fever has long attracted attention. It has been noted that rheumatic fever occurs chiefly in the first two decades of life while subacute bacterial endocarditis occurs more often after the second decade. (10) Saphir and Wile (56) from an analysis of clinical records, found that there is little significance in the number of preceding rheumatic attacks, the interval between the attacks, or the length of time elapsing from the last clinical manifestation to the onset of subacute bacterial endocarditis.

Fulton and Levine (25) found that in their series there was a striking freedom from recurrent rheumatic infections in the patients that developed this disease. Those patients who had frequent recurring bouts of rheumatic attacks did not usually succumb to this disease. It appeared to them, that those who became clinically immune to rheumatic
fever were the ones most apt to develop subacute bacterial endocarditis. While, those that continue with rheumatic manifestations are much less likely to develop this condition.

There are several quite generally accepted viewpoints; first that bacterial endocarditis generally occurs as a vegetative process engrafted on the site of an old rheumatic endocarditis; second, mitral stenosis is rarely the background for subacute bacterial endocarditis; and lastly, that it is a rare occurrence for severe congestive heart failure and auricular fibrillation to appear in the course of subacute bacterial endocarditis. (25) The grave prognosis with the development of this condition is well recognized and needs no comment.

MISCELLANEOUS - The last group consists of miscellaneous diseases to which rheumatic patients are just as susceptible as other individuals.

<table>
<thead>
<tr>
<th>Percent</th>
<th>Cause of Death</th>
<th>no.</th>
<th>avg. age</th>
<th>duration</th>
<th>symptoms</th>
<th>History</th>
<th>Rhythm</th>
</tr>
</thead>
<tbody>
<tr>
<td>33.1%</td>
<td>Congestive failure</td>
<td>49</td>
<td>40</td>
<td>4.2</td>
<td>17</td>
<td>24</td>
<td>4</td>
</tr>
<tr>
<td>23.0%</td>
<td>Acute rheumatic carditis</td>
<td>34</td>
<td>30</td>
<td>1.9</td>
<td>5</td>
<td>21</td>
<td>1</td>
</tr>
<tr>
<td>11.5%</td>
<td>Evboli and thrombi</td>
<td>17</td>
<td>40</td>
<td>3.8</td>
<td>5</td>
<td>11</td>
<td>0</td>
</tr>
<tr>
<td>29.5%</td>
<td>Subacute, Bect. Endo.</td>
<td>43</td>
<td>38</td>
<td>.5</td>
<td>24</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>3.4%</td>
<td>Misc.</td>
<td>5</td>
<td>33</td>
<td>9.2</td>
<td>0</td>
<td>3</td>
<td>0</td>
</tr>
</tbody>
</table>

AURICULAR FIBRILLATION - This arrhythmia is a rather prominent feature of rheumatic heart disease. This subject has been thoroughly studied. But only a few have studied it in relation to a specific heart condition such as the rheumatic heart. However, DeGraff and Lingg (21) have made
a thorough study of the influence that this arrhythmia plays on the course of rheumatic heart disease. They found that, out of 1633 rheumatic heart patients, 445 or 27.3% developed auricular fibrillation. Put of 644 patients that died 276 or 42.8% were known to have developed this condition. This latter percentage probably represents the true incidence since the whole course of these patients is known.

The type of valve lesion seems to have some influence on the incidence of auricular fibrillation. It would seem that mitral stenosis, or insufficiency, is necessary for its development. And auricular fibrillation rarely appears in the absence of these valvular defects. (22)

<table>
<thead>
<tr>
<th>Severe mitral stenosis</th>
<th>57% developed auricular fibrillation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>36%</td>
</tr>
<tr>
<td>&quot; insuff.</td>
<td>7%</td>
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<tr>
<td>No mitral involvement</td>
<td>0%</td>
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However, the the series is small, DeGraff and Lingg (21) found that one half of the cases with pure aortic disease of rheumatic origin developed auricular fibrillation.

This arrhythmia was found to be present in 57% of congestive failures; 35% in acute rheumatic carditis; 99% in those who died of emboli and only 2% in subacute bacterial endocarditis cases. (35) An apparent immunity to subacute bacterial endocarditis is well recognized. (22) (48) DeGraff and Lingg (21) found only 3 instances of this condition in their series of 644 patients who died with evidence of auricular fibrillation. The usual incidence in a group of heart cases is about 5%

They believe this is due to the fact that patients who develop subacute bacterial endocarditis have not had rheumatic heart disease long enough to have developed auricular fibrillation, since the latter is a late manifestation and the former occurs early in the course.

Patients can often name the exact date, and frequently the hour,
when auricular fibrillation began. It is usually present in the consistent type tho a few proxymal forms are noted. (21) The mean age at the time of development was found to be 37 years and the majority started between the ages of 27 to 45 years of age. This arrhythmia was most commonly observed in the 4th decade, consisting of 30.4% of the series. (21)

This arrhythmia rarely occurs in children, in 2539 cases of heart disease only 3 fibrillated. In another series of 1,164 there were no fibrillations. (48)

The age of rheumatic infection does not predispose to the onset of auricular fibrillation. Patients who were infected early, fell subject to this arrhythmia as often as those infected in later life. Except, when the initial infection occurred after 30, the incidence was higher. The duration of the disease and not the initial infection is the chief factor in its development. The patients who develop auricular fibrillation do not do so until after a mean duration of 18 years, in short, it is a late manifestation. (21)

In rheumatic heart disease, the cardiac reserve is frequently so
good that for several years patients present few or no symptoms. The onset of auricular fibrillation often being the first symptom prompting the patient to seek medical advice. On an average, this condition supervened four years after the symptoms of heart disease appeared. (21) It is the general impression that the duration of life after the onset of auricular fibrillation is often long. (12) (49) But other studies disprove this fact. The mean duration was found to be two years and two and one half to three years, in two different series. (20) (21) In many cases death took place within a year of the time that this arrhythmie began. This consisted of 34.3%. Seventy five percent died before three years had passed. Rarely, only 3 instances, did the patient for 10 to 12 years. For the most part auricular fibrillation tends to be a terminal event. It may be said that the chances of surviving the first year are 2 to 1 but the chances of surviving 3 years are only 1 in 4, and for 5 years only 1 in 9. (21) From controlled observations, it is apparent that the optimism as to the length of life after the onset of auricular fibrillation is unwarranted.

![Graph: Duration Life After Onset (20)]
The age of death on the average was higher when auricular fibrillation existed than when sinus rhythm was present, a mean of 36 years against 29 years. (21) Chapelle and Graeff give the average age at 43 years. Stroud et al. (65) gave 35.2 years as the average age of death. While there is some variation the general tendency of death to occur late in the course of rheumatic heart disease is evident.

In general whether patients with rheumatic heart develop fibrillation or not, they die of similar causes. (21) In about 68% congestive failure was the mode of death which is near that found in a group with normal rhythm. (22) The relation to subacute bacterial endocarditis has already been mentioned. Embolism and thrombosis caused death slightly more often among patients with auricular fibrillation, 11.5% against 8.4% in a group with normal rhythm. (21) This is contrary to the findings of Cookson (24).

The most important conclusion to be drawn, is that auricular fibrillation per se does not determine prognosis or life expectancy. This arrhythmia is a late manifestation of rheumatic heart disease and commonly observed in long standing cases. The die has already been cast when auricular fibrillation sets in. In other words, the heart damage is much more important than the arrhythmia. The longer the disease lasts the greater are the chances that a patient will develop this irregularity. This being the case, it is but natural that the average duration of life is short once auricular fibrillation is established.
SUMMARY

It is rather difficult to present a summary of this subject because of the varied findings and observation that are encountered. However, if one considers the general course and does not try to become too specific the course and prognosis is quite characteristic. The initial onset of rheumatic fever ranges from about 5 to 15 years of age. It can be assumed for practical purposes, that the heart is involved in almost all cases of rheumatic fever. However, the extent of involvement is quite variable. In many cases it will be difficult to tell if the heart is being affected at all. In other cases, there will be definite manifestations of carditis. The course will be more favorable if the heart is not enlarged and when a mitral murmur is soft in character. The presence of spontaneous nodules and pericardial friction rub give a much less favorable prognosis. Only a very few patients will die of the acute process in the first attack.

The future course of the patients that survive the first attack of rheumatic carditis is quite varied. The next three to five years represent the most important period. It is a safe rule that if the signs are going to disappear they will do so within a year from the onset. On the other hand, recurrences are almost certain to impress increasing disability upon the heart. As the months pass without recurrences the better the prognosis becomes. The individual prognosis depends upon the degree of infection and how much the heart is involved, as determined by symptoms and signs.

Following this dangerous early period the course splits depending upon the individual case. A certain number will present neither signs nor symptoms of cardiac involvement. Rarely these patients may live out their normal run of life without any disturbance. But more likely they will subsequently develop valvular lesions or will die of myocard-
ial failure from recurrences of activity. Another group will remain with an apical systolic murmur. If this murmur is faint, they will follow the course in the first group. If the murmur is loud in character, they are much more likely to develop mitral stenosis, though an exceptional case will live a normal life. The last group is that with evidence of some cardiac damage. There is some chance of them recovering but it is so small that one is not justified in holding out for it. There being only one chance in five that he will reach 20 years of life. And if this age is survived, there is only a maimed and crippled existence awaiting the patient. This of course depends again upon factors of reinfection and heart damage.

About one third will develop chronic valvular disease in the course of 10 to 15 years after the onset of the disease. While about 85% eventually will develop this defect, the development of mitral stenosis is by far the most likely. The future course of these patients is determined not so much by the type of valvular defect as by the degree of cardiac enlargement and the degree of cardiac failure. Many will live out all of 10 years uneventfully, some dying before this and a few living considerably longer.

On an average, patients will live about 11 years entirely free from symptoms. Then about the age of 28 there will appear symptoms of cardiac involvement consisting mainly of fatigue and dyspnea. This will culminate in the appearance of heart failure in about two years time. From this time to death, in about 3 more years, they are wholly invalids or at least in most cases seriously incapacitated. The average duration of life from the onset to death will have been approximately fifteen years. The age of death will be most likely from 23 to 42 with an average of 33 years. The cause of death is most likely to be congestive heart failure. The younger the patient the
more likely that death will be due to the presence of activity. In the older patients, it results from mechanical disturbance of the valvular deformity. As the decades pass there is increasing chance that auricular fibrillation will appear. At a certain age, about 50, the chances of this added burden are greater than the chance of escaping it. Other risks are thrombi with embolism, bacterial endocarditis and things that causes death in normal individuals may interrupt the course of the rheumatic heart case.
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