5-1-1940

Review of the literature on Pick's Disease

Robert E. Neurnberger
University of Nebraska Medical Center

This manuscript is historical in nature and may not reflect current medical research and practice. Search PubMed for current research.

Follow this and additional works at: https://digitalcommons.unmc.edu/mdtheses

Part of the Medical Education Commons

Recommended Citation

This Thesis is brought to you for free and open access by the Special Collections at DigitalCommons@UNMC. It has been accepted for inclusion in MD Theses by an authorized administrator of DigitalCommons@UNMC. For more information, please contact digitalcommons@unmc.edu.
A REVIEW OF THE LITERATURE ON PICKS' DISEASE

By
Robert E Nuernberger

SENIOR THESIS

Presented To The College Of Medicine,

University Of Nebraska, Omaha, 1940.
# TABLE OF CONTENTS

<table>
<thead>
<tr>
<th>CHAPTER</th>
<th>PAGE</th>
</tr>
</thead>
<tbody>
<tr>
<td>I- Introduction</td>
<td>1</td>
</tr>
<tr>
<td>II- Definition</td>
<td>4</td>
</tr>
<tr>
<td>III- Historical Resume</td>
<td>9</td>
</tr>
<tr>
<td>IV- Etiology</td>
<td>21</td>
</tr>
<tr>
<td>V- Diagnosis</td>
<td>23</td>
</tr>
<tr>
<td>VI- Differential Diagnosis</td>
<td>53</td>
</tr>
<tr>
<td>Mitral Stenosis</td>
<td>53</td>
</tr>
<tr>
<td>Polyserositis</td>
<td>55</td>
</tr>
<tr>
<td>Cirrhosis of the Liver</td>
<td>57</td>
</tr>
<tr>
<td>Nutritional Edema</td>
<td>58</td>
</tr>
<tr>
<td>Beri Beri Heart</td>
<td>58</td>
</tr>
<tr>
<td>VII- Physiology and Pathology</td>
<td>59</td>
</tr>
<tr>
<td>The Heart</td>
<td>59</td>
</tr>
<tr>
<td>Adhesions</td>
<td>63</td>
</tr>
<tr>
<td>Calcification</td>
<td>66</td>
</tr>
<tr>
<td>Paradoxical Pulse</td>
<td>69</td>
</tr>
<tr>
<td>Ascites without Edema</td>
<td>71</td>
</tr>
<tr>
<td>Liver</td>
<td>73</td>
</tr>
<tr>
<td>Polyserositis</td>
<td>75</td>
</tr>
<tr>
<td>Experimental</td>
<td>77</td>
</tr>
<tr>
<td>VIII- Treatment</td>
<td>79</td>
</tr>
<tr>
<td>IX- Prognosis</td>
<td>87</td>
</tr>
<tr>
<td>X- Presentation of Case</td>
<td>90</td>
</tr>
<tr>
<td>XI- Conclusions</td>
<td>104</td>
</tr>
</tbody>
</table>
INTRODUCTION

It matters not so much what one calls a condition so long as he knows that with which he is dealing and how to cope with it. Yet it makes a great deal of difference to the neophyte in search of information to find that what he has come to understand as a horse being called a cow. His whole perspective becomes changed and he is as it were in the middle ages when false premises were rife and Logic was King and the monks argued as to how many angels could dance on the point of a needle. Even so, eventually, if his interest does not wane he comes to understand that when some talk about something they mean something else-- and the trouble is that the foundations upon which knowledge is built is relative. The neophyte has gained some sort of an understanding of the subject--- he accepts the few men whom he can recognize as authorities --- and he is now able to give a book review, or write a paper to further befuddle the ones who will listen to him. Perhaps this is progress of a sort.

My interest in the subject of Picks' Disease was lighted by being present at autopsiy in a case which was diagnosed as such. I started out voraciously in my search of knowledge regarding this condition, and the more I read the more my reading danced to the tune of the Hindoo Fable regarding the blind men and the elephant, and so without excuses I present this poem as my introduction.

It was six men of Indostan
To learning much inclined
Who went to see the Elephant
(Though all of them were blind),
That each by observation
Might satisfy his mind
The First approached the Elephant,
And happening to fall
Against his broad and sturdy side,
At once began to bawl:
"God bless me! but the Elephant
Is very like a wall!"

The Second, feeling of the tusk,
Cried, "Ho, what have we here
So very round and smooth and sharp?
To me 'tis mighty clear
This wonder of an Elephant
Is very like a spear.

The Third approached the animal,
And happening to take
The quivering trunk within his hands
Thus boldly up and spake:
"I see," quoth he, "the Elephant
Is very like a snake!"

The Fourth reached out an eager hand,
And felt about the knee.
"What most this wondrous beast is like
Is mighty plain," quoth he;
"Tis clear enough the Elephant
Is very like a tree!"

The Fifth who chanced to touch the ear,
Said: "E'en the blindest man
Can tell what this resembles most;
Deny the fact who can,
This marvel of an Elephant
Is very like a fan!"

The Sixth no sooner had begun
About the beast to grope,
Than, seizing on the swinging tail
That fell within his scope,
"I see," quoth he, "the Elephant
Is very like a rope!"

And so these men of Indostan
Disputed loud and long
Each in his own opinion
Exceeding stiff and strong,
Though each was partly in the right
And all were in the wrong!
Men, in discussing what may or may not be Pick's syndrome, refer to the condition of Pick's Disease as concretio pericardi, synechiae pericardi, symphyses cardiacae, pericarditis chronicae adhesiva, callous pericarditis, chronic constrictive pericarditis, adhesive pericarditis, adherent pericardium, chronic mediastino-pericarditis, obliteratorive pericarditis, concretos disease, calcified pericardium, pericarditic pseudocirrhosis and so on ad infinitum.

Yet, a diagnosis of Pick's Disease is of value because

1. It affords the explanation of a group of symptoms and signs in an obscure case.

2. It obviates confusion with other conditions especially mitral stenosis, cirrhosis of the liver, polyserositis, nutritional and other rare forms of edema.

3. Expert thoracic surgery may now lead to cure of what once was a hopeless disease.
DEFINITION

In 1896 when Pick described three cases of constrictive pericarditis which presented an unusual syndrome--one of which was diagnosed ante-mortem--he brought to light a condition which had been known for a long time. Pick, however, was the first to associate the liver changes with the cardiac condition. In his description of the syndrome he informs us,

"There is a symptom complex of constrictive pseudocirrhosis of the liver which is deceptively similar to one of the mixed forms of hepatic cirrhosis with enlarged liver and considerable ascites but no jaundice. This pseudocirrhosis of the liver is caused by disturbances of the circulation of the liver due to latent pericarditis. These circulatory disturbances lead to an increase in connective tissue (fibrosis or cirrhosis) which in turn causes stasis in the portal circulation with marked ascites. This symptom-complex is found preponderantly in young individuals but may be observed also in after periods of life.

The following points are important in the differential diagnosis: a. Absence of an etiologic factor for cirrhosis of the liver. b. History of a previous pericarditis, c. Earlier occurrence of edema of the legs.

Certainty can come only through subsequent examination of the heart."

Freedman expresses the general present day opinion regarding the syndrome when he says,

"There is still considerable confusion concerning the nomenclature of this condition. 'Adhesive pericarditis' is not a correct term because the lesion is generally not an active inflammatory condition, but is the end result of a previous inflammatory process. Furthermore, the clinical syndrome, may occur without intra-pericardial adhesions and in turn intra-pericardial adhesions alone do not necessarily lead to the production of the syndrome. The term Pick's Disease has to be defined to be understood and, by itself gives no clue to the pathological anatomy or physiology of the disease."

Freedman is of the opinion that the term compression of the heart due to pericardial scar is much better than the term Pick's Disease. He does not believe that adhesions between pericardium and pleura or between the leaves of the pericardium are of much importance "since the clinical symptomatology of the condition is
not caused by the adhesions but by the pressure exerted by the thick pericardial scar on the heart."

Beck, in a discussion of the syndrome as regards physiology and pathology comes to the conclusion that,

"Anatomical terms, such as one encounters in the literature on disorder of the pericardium, are frequently devoid of clinical significance. This statement can be illustrated readily by a consideration of the most common of the pericardial lesions, namely, adhesive pericarditis. What does this anatomical condition signify? Does it mean that an inflammatory process must be present as the name implies or may it incorrectly refer to an adhesion or scar in which inflammation no longer exists? Are the adhesions intrapericardial or are they extra pericardial? Is there a disturbance of the circulation of the blood when adhesive pericarditis is present? If so, how is this disturbance brought about? Is it by a harness of adhesions upon which the heart tugs with each systole, or by the angulation of the heart away from its normal axis? In the treatment of adhesive pericarditis should pericardectomy or the Brauer operation be done? These questions adequately indicate the inadequate terminology of a condition labelled adhesive pericarditis. The disorders of the pericardium express themselves physiologically in one of two ways. They produce either the syndrome of acute intra-pericardial pressure or the syndrome of chronic intra-pericardial pressure. To understand these pressure syndromes, it must be remembered that under normal conditions the pressure in the pericardial cavity is negative. (The intra-pericardial pressure is approximately the same as the intra-pleural pressure. It is brought about in the same way as the intra-pleural pressure, namely, by the elastic recoil of the lung. The pressure of the venae cavae and in the right auricle is also negative (by about 3 to 4 cm. of water). Under normal conditions the circulation of blood meets no obstruction where the venae cavae enter the pericardial cavity, but under abnormal conditions a delicate and vital mechanism is called into play. If the pressure inside the pericardial cavity is increased, the venae cavae and right auricle are immediately collapsed and remain collapsed until the blood in the venous reservoirs assembles sufficient pressure to break through this intra-pericardial barrier. In this event the circulation starts up again. If the pressure in the venous reservoirs cannot build itself up to equal the pressure in the pericardial cavity, the circulation comes to a complete and permanent standstill. It is incompatible with life when the pressure in the pericardial cavity is greater than the pressure in the venae cavae. If the blood in the venous reservoirs can break through the intra-pericardial barrier, an adaptive mechanism takes place. If the intra-pericardial barrier is fluid or gas, the parietal pericardium will stretch in response to the pressure. However, if the parietal pericardium is thickened by the formation of scar tissue, it may be unable to stretch."

White and Churchill in an early article discuss the syndrome as being one form of chronic adhesive pericarditis. They list two forms, 1. cardiac adhesions to the chest wall or diaphragm, and 2. constricting adhesions over the heart chambers and great vessels. They believed that in the first case the reaction has been so extensive that it has spread beyond the visceral pericardium itself and has resulted during the process of repair, in a gluing of the heart to actively moving structures. These men believe that the second form differed from the first in that the handicap was not primarily one of increased work for the heart but was rather one of mechanical obstruction to the entrance of blood into the heart through compression of veins and heart chambers by thick, solid contracted membrane that encased them. Further, these men think that "the so-called chronic mediastino pericarditis, when it takes the serious course of Pick's Disease with evidence of obstruction to the circulation especially manifested by engorgement and eventual cirrhosis of the liver with ascites is but a form of chronic constrictive pericarditis. Chronic mediastinitis sufficient to obstruct the circulation directly is generally associated with constricting pericarditis which is the essential responsible factor for the Pick syndrome. Involvement of the anterior mediastinum with cardiac adhesions to the thoracic wall constitutes the other important type of adhesive pericarditis. The term mediastino pericarditis alone is insufficient designation."

Brooks and Lippencott, however, are of the opinion that chronic adhesive pericarditis although a lesion of great frequency is often impossible of diagnosis and in itself of very little clinical significance or importance. They do not believe that serious symptoms arise from the adhesive pericarditis unless the myocardium itself is seriously diseased.

Yater has the same idea when he says that chronic adhesive pericarditis practically never causes chronic cardiac compression.
Broadbent quite early describes several cases of adherent pericardium. One of these cases described, presented the typical Pick's syndrome. Many today, when they speak of adherent pericardium, speak of it in the same sense as Broadbent. To him the term implied the existence of adhesions between the visceral and parietal layers of the pericardium. He discussed only the universal type — that is, the cases in which the pericardium and heart were so intimately connected that the pericardial cavity was entirely obliterated.

Another condition is introduced by Sprague in his definition of the syndrome. He states that

"Chronic mediastino-pericarditis may produce to a unique degree a mechanical embarrassment of the heart such as Pick's syndrome. The degree of this is variable and would appear to depend for its gravity in the individual case upon the extent to which the adhesions encroach upon the superior and inferior cavae as well as upon the right auricle and other heart chambers. Whether the picture as presented by the patient is that of congested head, neck, and thoracic veins (superior mediastinal syndrome) or of enlarged liver, ascites, and portal engorgement (inferior mediastinal syndrome) is, in turn dependent upon the relatively greater involvement of the superior or inferior cava. In most cases both great veins are affected either primarily, or more often secondarily, through interference to diastolic filling by constitution of the heart from a thickened or calcified visceral pericardium.

Assuming that the diagnosis of obstruction to the inflow of blood to the right heart by pericardial adhesions can be made, a subject beyond the purpose of this paper the relief of this obstruction should be considered."

The best definition of the syndrome is that given by White who considers chronic constrictive pericarditis and Pick's Disease as synonymous. To quote,

"Chronic constrictive pericarditis consists of a chronic fibrous or callous thickening of the wall of the pericardial sac which is so contracted that the normal diastolic filling of the heart is prevented. A condition called inflow stasis results. There may or may not be calcification of the pericardium, obliteration of the pericardial cavity or important external pericardial adhesions. There may or may not be an associated accumulation of pericardial fluid in small amounts, as in pockets. The parietal pericardium may be pre-
ponderantly affected or the epicardium also seriously involved; or both pericardial membranes may be securely or even inseparably united. One section of the pericardium as over the cardiac apex, may remain relatively free and but slightly thickened while another part, as over the right auricle and great veins is markedly contracted, or the entire heart and roots of the great vessels may be encased uniformly in a tightly fitting envelope.

There may or may not be an acute or chronic polyserositis, which is, contrary to a common belief, a different thing. There may or may not be a frosted liver or spleen. There may or may not be heart disease itself -- in my experience the association of chronic constrictive pericarditis of any important degree at least, with heart disease is very rare; I have encountered only one likely case, and this I believe to be a coincidence.

The signs of chronic constrictive pericarditis are similar in most respects to those of acute constrictive pericarditis, or cardiac tamponade, due to pericardial effusion and indeed the signs of the former condition may follow the signs of the latter without any interruption at all, or more often after any interval of months or years. The insidious evolution of the disability due to chronic constrictive pericarditis, frequently with no history of a preceding acute pericarditis, makes the diagnosis far more difficult than that of acute constrictive pericarditis that usually develops with striking signs and symptoms in the course of a few days or hours."

In order to show the state of our present day knowledge regarding Pick's syndrome, quotations from two of the outstanding surgeons who have dealt with this disease will suffice.

Beck \(^{11}\) recommends the use of the term pericardial scar instead of adhesive pericarditis or adherent pericardium. He does this because he believes that the term does not indicate the presence or absence of adhesions. In another article Beck \(^{12}\) says,

"I feel secure in stating that adhesions to the heart do not produce dilatation, failure, or hypertrophy of the heart. Adhesions play no part in the production of the compression syndromes. When present, adhesions are silent and incidental findings and produce no circulatory trouble whatsoever unless the heart is acutely un- gulated or twisted. Experimental and clinical evidence conclusively leave no room for doubt."

Churchill, \(^{12}\) completing our confusion states that "We ordinarily think of Pick's syndrome as originating from a completely encased heart, but as time goes on we are accumulating evidence about the effect of localized pericardial adhesions."
HISTORICAL RESUME

Diseases of the pericardium are among "the most venerable
of recognized cardiac lesions in the history of medicine." 6
"Galen, about 160 A.D. described pericardial effusion in a monkey
and scirrhous thickening of the pericardium in a cock and surmised
that the same conditions might occur in man and interfere with
proper cardiac function." 10 Galen also successfully operated on
a young man, removing some of the diseased sternum and even the
vertex of the pericardium which he uncovered in a case of post
traumatic septic anterior mediastinitis. This is the first record
of pericardial resection. 10 Galen, however along with most of the
early writers believed that in cases of adherent pericardium there
"was congenital absence of the pericardial sac." 3

"Among the ancients the 'hairy heart' was supposed, according
to Haller to indicate great valor and bravery. Among those heroes
who after death were found to be so endowed are said to have been
Leonidas and Lysander." 6

"Gentilis of Fuligno, 1518, attributed this condition to
hypertrophy of the pericardium, accompanied by atrophy of the
heart."

"Boerhave, in his writing, collected and published by Haller,
mentions adherent pericardium as a cause of palpitation and cardiac
distress leading subsequently to death. The adherence of peri-
cardium he attributes to deficiency of 'vapor' in the pericardial
cavity......

"Rondelet described the pain, dyspnea and syncope of pericar-
ditis and Riolan, in 1649 recognized the danger of the condition
and boldly suggested incision and drainage of the pericardium
after trephining the sternum." 14

Lower, 1669, should receive credit for the first satisfactory account of pericardial disease in man. He wrote that "a profuse effusion oppresses and inundates the heart.... The walls of the heart are compressed by the fluid settling everywhere so that they cannot dilate sufficiently to receive blood; then the pulse becomes exceedingly small, until finally it becomes utterly suppressed by the great inundation of fluid, whence succeed syncope and death itself." 15 He also recognized that interference with the proper heart action could arise when there are thick contracting adhesions. He cited a case of a London housewife, age 30, whose pericardium "was not, as is proper, thin and transparent, but thick, opaque and even callous," 16 and introduced the term "callous pericarditis."

Vieussens, 1715, cites a case of adherent pericardium, in a child of five, and in which the pericardium was found to be thick and firm, and almost cartilagenous in nature. He mentions among the symptoms "dyspnea which he attributes to the inability of the diaphragm to descend perfectly in inspiration, owing to its being bound down to the heart by pericardial adhesions." 8

Lancisi, in 1728, made the clinical picture of chronic constrictive pericarditis by describing the small pulse, engorgement of the jugular veins, and enlargement of the abdomen in a young man in whom necropsy revealed a small heart with thick completely adherent pericardium. 10

To complete the first century of the recognition of chronic adhesive pericarditis, Morgagni 8 in 1761 discussed very fully the question of adherent pericardium. He states that many writers had
mistaken cases of adherent pericardium for absence of pericardium to refute this idea. He gives an account of 45 cases of adherent pericardium, collected from the writings of various authors, whose names he gives, and he describes the main features of each case. He states that palpitation had been described as a prominent symptom of adherent pericardium by Vieussens and other writers, but this he does not regard as a characteristic symptom; it was present in only 15 out of the 45 cases described. Absence of or weakness of the apex he considers an important physical sign; from an analysis of these cases, finding it 30 times.... He mentions smallness and irregularity of pulse, palpitation, shortness of breath, oedema and ascites as some of the symptoms." 8

Morgani also reported seven cases, one of whom, "a man 40 years old with a very small pulse, showed a heart 'so constricted and confined that he could not receive a proper quantity of blood to pass on.' But he noted that most of the cases were incommended little or not at all even shortly before death." 8

With this idea of the uncertainty of making the correct diagnosis which was reiterated by Corvisart in 1806, 17 and Laenner in 1819, 10 the attention of the medical profession in general was distracted from the possible importance of chronic constrictive pericarditis.

The idea of operating on the heart for relief certainly didn't enter many heads during this period. It is true that Senac 14 about 1749 advised incision and drainage for pericarditis describing a method. However, the heart was held to be sacred and not to be touched. This attitude was as heritage from the past. Hippocrates,
Aristotle, Pliny, Fallopius, and other imposing names gave teeth to the tradition by stating that all wounds of the heart were necessarily fatal, and that if the heart were wounded it would never heal, "being too hard in motion and of an inflammatory heat."\textsuperscript{14}

Owing to the tradition and the difficulty of making a correct diagnosis only two attempts were made to deal with the pericardial cavity. In 1798 Desault incised and drained an incised pleurisy. In 1819 Romero performed the first successful pericardiotomy. In operations on three cases of pericarditis with effusion, two of the patients recovered. A puncture of a distended pericardium was made by Jowett in 1827. No other cases except an attempt by Schick to tap the pericardial cavity are recorded until 1841 when a great number of successful cases was reported by Karanoeff following an outbreak of scurvy in Cronstadt Russia.

In the year 1842 Chevers\textsuperscript{18} from Guy's Hospital gives us our first clear statement and understanding of constriction of the heart. He says,

"The principal cause of dangerous symptoms appears to arise from the occurrence of gradual contraction in the layer of adhesive matter which has been deposited around the heart, compressing its muscular tissue, and embarrasing its systolic and diastolic movements but more particularly the latter. Under these circumstances, the circulation seems, after a time, in great measure to adapt itself to the encumbered condition of the heart. The ventricles, having become diminished in capacity, make up for this loss by the rapidity of their contraction (hence the small and rapid pulse, noticed in the above case); while the main arteries if not already diseased, adapt themselves to the dimensions of the cavities from which they arise. And thus the blood passes onward for a time, with tolerable freedom; but the patients become incapable of continued muscular exertion, and are always liable to suffer from dropsy and other serous effusions upon the occurrence of very slight pulmonary obstructions. In the case which I have quoted, the serous effusions which gave rise to the most prominent symptoms of disease evidently arose from the cavities of the heart being no longer capable of transmitting the blood with ordinary freedom. The heart had, doubtless, for a long time continued to
become more and more compressed, weakened and embarrassed by the gradual contraction of the adventitious structure which surrounded it; distention of the great veins and abdominal viscera had necessarily followed; and the resulting anaemia and ascites must have added still more to the obstruction with which the already powerless heart had to contend. The patient died from the effusion of serum into the medullary substance of the brain (that organ, having been rendered susceptible by previous disease). In the other case it does not appear that the shackleing of the heart had gone on to a degree sufficient to produce any serious result."

Dr. Hope, however, the authority of that time had observed that he had never examined, after death,"a case of complete adhesion of the pericardium without finding enlargement of the heart, generally hypertrophy with dilatation," but added that the adhesions may not for a time produce much inconvenience. It was his opinion that in such cases, where the adhesions are close and universal, the incessant struggle over distention of the heart, together with inflammatory action, tends invariably to cause enlargement of the organ.

Dr. Chevers, however, says,

"With deference to this high authority, I must venture to suggest that the above remarks can be applied only to one class of cases of this description — to those in which, superadded to the adhesion of the pericardium, there is also disease of the valvular passages of the heart: these cases are certainly the most frequent; but I think that a sufficient number of contrary instances have occurred, under my notice, to prove, that where the valves are healthy, complete and close adhesion of the pericardial surfaces, far from producing hypertrophy and dilatation, has a tendency to be followed by general diminution in the size of the heart and its vessels, and contraction of its cavities....

"I believe I may confidently state, chiefly from my own observations, and also from a careful survey of the Museum Records, that during the last seven years, no case of decided hypertrophy with dilatation of the heart, occurring in combination with long standing obliteration of the pericardial cavity has been inspected at Guy's Hospital, in which there was not also present either morbid narrowing of the aortic or mitral orifice, or some evident obstruction to the pulmonary or systemic arteries, causes fully sufficient in themselves to produce enlargement of the heart cavities in each case, quite independently of the adherent state of the pericardium."
Although Chevers wrote in the great days of Bright and Addison his observations escaped unnoticed.

Certainly the pathology of an encased heart -- in particular a calcified one was not unknown to the American nation in this period. In the year 1851 N. Hawthorne published Ethan Brand. Hewitt has called our attention to this story which contains a vivid description of the death of the main character who had a 'heart of stone'.

Ethan Brand is related to have committed suicide by plunging into the burning kiln which in former days he had tended. Hawthorne concludes the story as follows,

"With his long pole in his hand he (Bartiam) ascended to the top of the kiln. After a moment pause he called to his son. "Come up here, Joe!,' he said. So little Joe ran up the hillock and stood by his father's side. The marble was all burnt into perfect snow-white lime. But on its surface, in the midst of the circle, snow-white, too, and thoroughly converted into lime, lay a human skeleton, in the attitude of a person, who, after a long toil, lies down to long repose. Within the ribs, strange to say was the shape of a human heart.

"Was the fellow's heart made of marble?" cried Bartiam, in some perplexity at this phenomenon.

"At any rate, it is burnt into what looks like special good lime; and taking all the bones together, my kiln is half a bushel the richer for him."

"So saying the rude lime-burner lifted his pole, and letting it fall upon the skeleton the relics of Ethan Brand were crumbled into fragments."

In 1855 Stokes, writing of adherent pericardium says,

"Without denying that a general adhesion may induce hypertrophy and dilatation, experience leads me to doubt that such an effect necessarily or even commonly follows the condition indicated. I have often found the heart in a perfectly natural condition, with the exception, of an obliterated pericardium. It was neither hypertrophied nor atrophied, and the patient had exhibited no symptoms of heart disease for many years before death. In one case seven years had elapsed between the death of the patient from hepatic disease, and the attack of pericarditis which obliterated the sac... It is in those cases of pericarditis which we have before indicated, and where valvular disease is either co-existent with or
or subsequent to the first inflammation of the sac, that hypertrophy and dilatation appear as remote consequences of pericarditis...

It has been stated to me by Professor Smith that he has found general adhesions of the pericardium coinciding with atrophy or with hypertrophy of the heart in a nearly equal frequency.

In one, as in adhesion, the normal condition of the muscle is interfered with, and so that contraction diminished; while in the other the muscle (mitral disease) being free to act, increases in power.

On the whole we may conclude:

1. That obliteration of the pericardium does not necessarily endure any manifest changes in the condition of the heart.

2. That, when alteration of the muscular condition of the heart is found in connection with this obliteration, it is not necessarily a state of hypertrophy, but is often one of an opposite nature.

3. That the cases of valvular obstruction and of adhesion of the pericardium are not parallel, in as much as that in one case the heart is free to act while in the other its motions are prevented or interfered with.

4. That obliteration of other serous membranes is more often followed by atrophy than by hypertrophy of the subjacent organs.

In 1871 Wilks, another Guy's Hospital man calls our attention to Chever's article and after presenting 6 cases, one a boy of 9 years, who had Scarlatina 3 years prior to his illness, he writes,

"It may be that the position of the adhesions may cause a difference in this respect, and thus the result vary according as one portion or another is bound tight. As a rule, the greatest thickening is in front, so as to affect the right auricle and ventricle.

I would especially draw attention not only to the fact that the chronic inflammatory process very often involves the vessels entering the heart within the pericardium, but that when the latter is adherent the thickening and induration proceeds, as would a growth, to involve the cellular tissue in the mediastinum, and even creeping upwards, to surround the veins in the neck. I have thus seen, in two or three cases, a tough areolar tissue surrounding the vena cavae and brachio-cephalic veins in connection with an old pericarditis...."

But even at the present day the question is still asked, are pericardial adhesions of any importance or not? The answer probably is, that loose cellular adhesions have no appreciable influence on the action of the heart, but that the thickened pericardium of a cartilaginous consistency investing the heart closely, arising from an inflammation at an early period of childhood, does lead to ob-
struction of circulation, and then to dropsy, after the ordinary manner of heart disease.

Along with these two Englishmen, two Germans, Greisinger and Kussmaul, 1873, called to the attention of the medical world, the syndrome of chronic constrictive pericarditis. Kussmaul, under the title of "schwielige (callous) pericarditis" with Griesinger’s observations in mind made ante mortem the correct diagnosis in two cases with this syndrome.

However, despite the observations little attention was paid to chronic constrictive pericarditis until after Pick’s contribution in 1896. Incidental papers were written by Concato on polyserositis in 1881, by Hitinel on "cardiac and tuberculous cirrhosis of the liver,"in childhood and by Harris in indurative mediastino pericarditis in 1895.10

Broadbent in 1895 published a monograph on adherent pericardium. From his definition of his title it is possible he might have had chronic constrictive pericarditis in mind -- but all except one of the cases which appears to be chronic constrictive pericarditis -- the others are cases of adherent pericardium.

From the standpoint of surgery in treatment of chronic constrictive pericarditis matters were even more at a standstill. In 1870 Billroth characterized the operation of tapping the pericardium "as a surgical frivolity and a prostitution of surgical skill." And again in 1883 declared that no surgeon who wished to preserve the respect of his colleagues could ever attempt the suture of a wound of the heart.14 This was the way matters stood when Pick in 1896 submitted his paper entitled "concerning chronic pericarditis
running its course under the guise of cirrhosis of the liver
(pericarditic pseudo cirrhosis of the liver) with observations on
the frosted liver. (Curschman).

In this same period Weill in 1895 suggested that relief might
be brought to these patients some day by pericardial resection.

Churchill quotes him as saying "After the adhesions have
reached a fibrous stage, they act independently of their original
cause, and medical therapy is illusory. One has never dreamed of
attempting a debridement in such a case, less perhaps from resig-
nation than from the uncertainty of the diagnosis. There would
be good cause to make an attempt of this kind, designed to liberate
a part of the heart, the apex for example, and the anterior sur-
face.... It will some day come within the provinces of surgery to
deliver the heart from the shell which strangles it."

To the French Surgeon Delorome in 1898 goes the palm of dis-
tinction for insisting that an operation might be done for chronic
constrictive pericarditis in which the thickened pericardium is
divided and stripped from the heart muscle. He stated in detail
what the operation should be from observations of cadavers. He
did not do the operation. His suggestions were not followed,
however, until in 1913 when Rehn and Sauerbruch independently did the
operation. Rehn at this time compared the constricted heart
to the constriction of the hand by a tightly fitting glove.

It is of interest to close the 19th century with contributions
from America. In 1898 Cabot reported a case of Pick's Disease
and collected 17 others from the literature. The case which Dr.
Cabot presented is of interest in that the boy who was 13 years old,
had diarrhea -- 3 to 5 movements daily, despite efforts to check
during the 8 weeks time that he was in the hospital. In the 18
cases which Cabot collected he comes to the conclusion that "the
absolute latency of the pericarditis is important but not on the
whole surprising." He watched a number of cases of pneumonitis and was especially on the lookout for the advent of pericarditis and examined the cases each day in vain "yet at autopsy the pericarditis was found to have crept in." Regarding the symptoms he states

"That weakness of the heart brought on by its adhesions to the pericardium should produce hepatic stasis and ascites without edema of the legs or anasarca is strange, but not at all without precedent in literature and experience... The most acceptable explanation of this special localization of the passive congestion in the portal system is that the stasis in the liver gives rise to permanent tissue change there, namely an increase of the stroma with contraction, over which the influence of rest and drugs has no effect. Other manifestations of passive congestion can be removed by treatment, but in the liver they are self perpetuating and grow in a vicious circle."

In diagnosing the syndrome, he was of the opinion that any unexplained enlargement of the liver with portal congestion and ascites should be considered with regard to the possibility as being due to adhesive pericarditis and goes on to state, "As most cases of chronic pericarditis are tubercular in origin, the presence of tuberculosis elsewhere in the body in connection with unexplained portal stasis should make us think of the pericardium as a possible cause for the stasis."

In 1901 in a discussion on adherent pericardium Dr. Carl Beck\(^27\) raises the question for the first time in America\(^24\) of the possibility of a surgical cure. He said,

"I should like to ask Dr. Babcock as to the advisability, in cases of adherent pericarditis of approaching the adhesions with the surgeon's knife. We are no longer afraid of the pericardium. I had great pleasure in seeing a boy, a few months ago, recover after opening and draining the pericardial cavity. Just as 25 years ago we were afraid to open the pericardial cavity, so now do we seem afraid to open the pericardial cavity, and it may be, the work of the surgeon in the treatment of pericardial effusions and their consequences will be somewhat similar to what it has been in pleuritis."

Turk, in 1901 wrote a paper on the differential diagnosis of
pericardial concretion. His conclusions were

1. Along with internal adhesions there may or may not be any important external adhesions with signs thereof, such as systolic retraction of the chest wall.

2. That polyserositis or peri-visceritis may antedate adhesive pericarditis.

3. Differentiation from TB peritonitis and from tricuspid and mitral valve disease is more difficult than from true cirrhosis, in contradistinction to Pick's point of view. He noted that the peritoneum may or may not be involved in these cases.

In 1902 the Brauer operation was introduced as a singular procedure for relief of chronic constrictive pericarditis. Generally good results were reported but at present it is regarded as just a method of approach to the heart for decortication.

In 1912 Dr. Summers performed the first operation of the Brauer type done in America. He believed, however, that the process of respiration profited much more by the operation than the heart action.

Our next step in the history of chronic constrictive pericarditis is Neils Finsen's account of the disease. He, himself, had the disease and treated it medically with low fluid intake and low salt intake for 21 years. Finsen, who won the Nobel prize in 1903 gives an interesting account of the symptoms as he observed them.

Following Rehn and Sauerbruch, Schmeiden in 1918, from a surgical standpoint and Volhard in 1923, from a clinical standpoint contributed important and classical papers regarding constrictive pericarditis.
After interest was aroused in chronic constrictive pericarditis a diagnostic aid by means of the X-ray developed. The possibility of discovering pericardial calcification roentgenologically was suggested by Diemer in 1899, and by Simmonds and by Lydten in 1907. It remained for Schwarz of Vienna to realize this possibility in 1910, when he reported a case in which fluoroscopy of the thorax revealed within the heart outline. 31

In 1924 Scholz 32 presents the first case diagnosed by X-ray and confirmed by autopsy.

The contributions since 1920 have largely been the work of White and Churchill, Blalock and Burwell, Beck and Griswold, and Cutler.
ETIOLOGY

There has appeared in the 44 years since Pick's report an extensive literature on the subject and one of the chief problems discussed has been the etiology of the condition.

The etiology of Pick's Disease is not known. It is certain that there is not just one factor and it is generally agreed that rheumatic fever is not the cause of the thick callous scar of chronic constrictive pericarditis.

We do not have much proof to bear out this statement except that the best men who have diagnosed the condition and have followed up by operations and autopsies have not found it to be so. The only research done along this line has been the study of 1000 children at the House of Good Samaritan in Boston who had Rheumatic fever and were followed for 10 years. There was not one instance of chronic constrictive pericarditis even though the heart was seriously involved and even though acute pericarditis had been noted in many cases during their acute rheumatic infection.

There are many men, however, who do not agree with this statement that rheumatic fever does not cause Pick's Disease. The question is certainly controversial when one considers that rheumatic fever is the chief cause of adhesive pericarditis. Rothstein regards rheumatism as the etiologic factor in chronic adhesive pericarditis and Pick's Disease. He regards Pick's Disease as merely one of the resultant symptom complexes which may result from any acute pericardial inflammatory processes. Howard states that "chronic or indurative mediastinitis usually follows rheumatic pericarditis, but may also develop subsequently to fibrinous pleurisy, or in association with some other of the acute conditions of the chest."
The biggest difficulty in ascribing etiologic factors of this syndrome is that our understanding of the pathology of the condition is incomplete. Tice informs us that Pick's Disease may occur either as a primary clinical entity or secondary to some other disease. He does not believe that the clinical symptoms and signs are due to the obliterated sac alone but result from the associating pathology, "the symptom complex of which is sufficient to give it special consideration and by which an adherent pericardium is recognized. He believes that every type of infection or disease capable or producing an inflammatory involvement of the pericardium must be considered as an etiologic factor. Tice divides adherent pericardium into 1. the silent group, 2. those presenting symptoms of chronic heart disease, and 3. those with Pick's syndrome. He feels that the usual manifestations of adhesive pericarditis are those of a chronic heart disease not due primarily to the adhesions but to the associated cardiac pathology.

Lyter in a discussion along similar lines states that from the standpoint of etiology pericardial adhesions fall into 3 classes.

1. The tuberculous,

2. Those resulting from an attack of acute pericarditis or more probably pancarditis secondary to or complicating some acute infectious disease as rheumatic fever, tonsillitis, lobar pneumonia, typhoid etc.

3. Cases which give no history of previous acute infections of any kind.... The adhesions in this class are probably the result of long and continued injury to the pericardium from chronic infections or some unknown intoxication. In this class we see most
typically the assorted chronic changes in the liver and kidney

(advanced Picks' Disease)

Lyter thinks that the pericardial, hepatic and peritoneal findings in Picks' Disease are quite probably not independent and distinct, but the result of a common etiology as chronic infection or intoxication.

Tuberculosis appears as an outstanding etiologic factor in many of the cases diagnosed as Picks' Disease. Churchill presents the present state of our knowledge regarding this factor well when he says

"The probability that many of the cases of chronic constrictive pericarditis represent healed stages of a pericarditis due primarily to tuberculosis is an important question still to be settled. Examination of the scar tissue removed at operation universally fails to establish a diagnosis of tuberculosis. On the other hand the frequency with which calcium deposits are found, and the unusual density of the scar tend to link the pathology with the tubercle bacillus.

The only reliable finding that distinguishes the active phase of tuberculous pericarditis from constrictive pericarditis due to a healed scar is the demonstration of the tubercle bacillus in fluid aspirated from the pericardium.

The sudden appearance of the syndrome in a patient with known active TB elsewhere is presumptive evidence.

Fever, increasing severe tamponade, bloody pericardial fluid, rapid sedimentation rate and other signs of active infection are suggestive, but are occasionally met with in patients not showing pericardial tuberculosis. In cases of reasonable doubt, it is best to make a limited exploration under local anesthesia. Active infection can usually be recognized by the presence of tubercles on the inner surface of the parietal pericardium.

Blalock found the tubercle bacillus to be the etiologic agent in most of the 22 cases he has seen. In discussing the etiology he states that infections of the tubercle bacillus and staphlococcus aureus "are particularly apt to result in chronic constrictive pericarditis, with compression of the heart. In fact all pyogenic infections of the cavity which are not drained are apt
to result in this condition. Following drainage of the pericardium for a pyogenic infection, chronic pericarditis has resulted once in the experience of my co-workers and me and twice in that of Bigger.

Sprague\textsuperscript{41}, in discussing Pick's Disease states that it is "a condition that is usually progressive from the time of the original infection, and the inflamed pericardium in passing into a sub acute stage or in healing gradually thickens as it enfolds the heart in its inelastic grip. Its etiology is unknown, but in our experience it would appear to arise from a tuberculous infection or from the agent responsible for respiratory infections, such as influenza and pneumonia. Rarely it follows septic pericarditis."

Reed,\textsuperscript{45} Wenckebach,\textsuperscript{46} Burwell,\textsuperscript{47} Rolleston,\textsuperscript{48} Konstam and Wood,\textsuperscript{38} Tannebaum\textsuperscript{56}, consider tuberculosis as a common etiologic factor in Pick's Disease. Ramsey\textsuperscript{49} suggests an avian tubercle bacillus as the cause.

Riesman,\textsuperscript{50} however, believes that tuberculous pericarditis is quite rare.

Maclachlan\textsuperscript{51} presents an unusual view regarding tuberculosis as an etiologic agent when he says,

"It seems rather strange that calcification, a cardinal sign of healed tuberculosis, when it occurs in the pericardium very rarely means this disease.

Certainly it is my impression that the tuberculous form of pericarditis which produces a recognizable clinical entity as a disease with a high mortality.

The pericardium, situated as it is in proximity to the common site of primary foci of tuberculosis in the thorax, does not present evidences of this disease as often as one would expect. The finding of a typical tuberculous lesion in the pericardium at autopsy is not rare, but, on the other hand, it is by no means common: while the demonstration of a recognizable clinical tuberculosis of this sac, is, to say the least, infrequent. On the other hand, there is little doubt that many of the clinical types of the disease are not diagnosed, and from the pathological aspect there is a considerable group of old healed fibrous lesions found at autopsy which are at times not unlikely tuberculous, but as to their exact nature it is a most difficult matter on which to speak positively."
Mitchell believes that tuberculous pericarditis may appear at any age, although it occurs more often in children and young adults. He says that it may complicate active TB elsewhere in the body (including miliary) or may be the only manifestation of early disease. He believes that it is seldom seen in chronic pulmonary tuberculosis but is more apt to appear in tuberculosis of the lymphatic system or the serous membranes. The disease, he feels, tends definitely to progress to a chronic, fibrous state in which thickening of the epicardium and pericardium and the development of adhesions become conspicuous features. These adhesions, he feels, tend to contract and thus to limit the action of the heart and obstruct, in varying degree, the flow of blood into this organ. Referring directly to Pick's syndrome he says,

"It is interesting to speculate upon the etiology of Pick's Disease, or at least upon the polycerotic aspects of the syndrome. Does effusion appear in the pleural and peritoneal sacs of people afflicted with Pick's solely or chiefly as the result of a failing circulation produced by pre-existing adhesive pericarditis or does the effusion represent response to specific local disease? We have seen a case of pulmonary TB complicated by what was believed to be TB peritonitis, the abdominal fluid being plus for TB bacilli; in which a perfectly clear peritoneal surface was found at autopsy several years later."

McLester believes that tuberculosis is of importance because the sclerotic process which accompanies it tends to involve the lower and posterior half of the mediastinum.

Barnes and Harrington in a discussion of the etiology state that

"Repeated attacks of pneumonia with pleuritic complications appear fairly frequently in the histories of these patients and one can readily believe that pericardial infection had its inception in one of these attacks. At present one must assume that the organism which invades the pericardium is of low virulence and that the patient can live without subjective awareness that the process is under way until the mechanical effects of inflow stasis produce clinical symptoms and findings."
Wells\(^54\) also believes that a large number of these cases are
due to pneumococcal infections.

We must not forget that anything which causes acute pericarditis may be responsible for the chronic form and so Pick's Disease.\(^{57} 58 59\) Gonorrhea, pneumonia, septicemia, erysipelas, pericarditis, and osteomyelitis, scarlet fever, typhoid, B. coli, measles, small pox, mumps and whooping cough, diphtheria, cerebrospinal fever, actinomycosis, scurvy and syphilis may all cause acute pericarditis and so must be considered as etiological factors of Pick's Disease.\(^{57} 60\) Branch\(^61\) states that the reason the cause of chronic constrictive pericarditis is so frequently unknown is because the acute condition escaped notice at the time.

Howard\(^62\) informs us that the chronic form of pericarditis may follow:"even an acute general infection, as scarlet fever, or measles, without the history of any recognized pericarditis or pleurisy. Occasionally it is chronic from the outset, secondary to a tuberculous focus of the mediastinal glands or of the adjacent lungs. TB is certainly a more frequent etiological factor than has been generally recognized. Syphilis, too, has played an undoubted role in a small proportion of the cases."

Infection of the pericardium may occur:

1. Through the blood stream.
2. Through the lymph vessels.
3. Direct infection by trauma.
4. By extension which may be from
   a. heart or vessels within the pericardium
   b. mediastinum
   c. pleura
   d. peritoneum

\(^54\) Wells
\(^57\) 58 59
\(^57\) 60
\(^61\) Branch
\(^62\) Howard
Stone states that the pericardium lies in intimate contact with the structures of the mediastinum and involvement of these structures occurs not infrequently during the course of infections such as pneumonia with empyema, rheumatic fever, scarlet fever, pyemia, and tuberculous pleurisy with effusion.

Lenker thinks that Scarlet fever might be the cause of Pick's Disease. Sprague believes that the inflammatory polyserositis which occurred in some patients during the influenza epidemic of 1918-1919 may possibly have been responsible for the latter development of constricting pericarditis in a few of our cases.

White, in a discussion of 15 cases of Pick's Disease which he presented found

a. Tuberculosis to be the cause in 2 cases, questionable in 2 others.

b. Pneumonia with polyserositis (including pleuritis and pericarditis in 2.

c. Sepsis in one case.

d. Rheumatism in none.

e. Uncertain or unknown in 10 although in 5 however there was a definite history of acute pericarditis.

Hunter and East in a consideration of three cases of chronic constrictive pericarditis were of the opinion that tuberculosis was the possible cause in two cases. Piersol, Griffith et al. believe Pick's syndrome to be caused in some cases by a low grade streptococcus of unknown origin. Borchart advises that a search be made pre and post operatively for foci of infection especially tonsils and teeth.

Fenton believes the condition to be the result of a chronic
degenerative factor, possibly dependent upon the presence of some toxic material, but, at the same time, is not especially associated with other well recognized degenerations of the renal or circulatory systems.

Willius presents a case of Pick's Disease which he considers scarlet fever as being the cause.

It would seem from a review of the literature that most anything which causes an acute pericarditis may be responsible for Pick's Disease. Tuberculosis and pneumonia appear to be responsible for the majority of the cases where the etiology is known. The reason that rheumatic infections do not result in a constrictive pericarditis is that there is not the long continued insult such as occurs, for instance, in tuberculous infections.
DIAGNOSIS

Sprague \(^4\) thinks that the diagnosis in this condition is not made because "there still exists in the minds of many physicians the belief that the diagnosis of chronic constrictive pericarditis is almost impossible and that there is little to be gained from it except a sense of diagnostic satisfaction on the part of the doctor."

Smith and Willius \(^6\) state that the didactic overemphasis on so-called characteristic signs has gone far toward frustrating correct diagnosis, for in reality such signs exist in relatively few cases, and their absence often influences the clinician against committing himself.

White \(^7\) states that in 80% of the cases, the diagnosis of adherent pericardium can not be made and that in these cases the adhesions are of no significance. However, in the case of the 20% of important pericardial adhesions he is of the opinion that a diagnosis should be made and in addition the differential diagnosis between the constrictive type and the other should also always be attempted.

Wusser and Herrmann \(^17\) analyzed cases of pericarditis in the Charity Hospital in New Orleans over a period of 5 years. They found that constrictive pericarditis is seldom recognized clinically.

To quote:

"Inflammation of the pericardium, be it acute or a chronic process, represents one of the pathologic lesions of the body which the clinician rarely diagnoses and which is discovered only at autopsy. The explanation of this observation lies not so much in the want of clinical data that may have a bearing on the diagnosis, but rather on the failure of the physician to make use of all the available facts disclosed by complete and thorough examination of the patient. The so-called pathognomic signs are looked for, and, if not observed, the case is dismissed as one without pericardial involvement. Conclusive signs are rare."
Sprague in listing the diagnostic criteria gives the following chief points:

1. Patient usually young child or young adult.

2. Ascites and enlarged liver appearing insidiously are always present and usually out of proportion to the peripheral edema.

3. The veins in the neck are engorged and the venous pressure in arms and legs is often over three times normal. Moreover the venous pressure remains constantly elevated, not fluctuating through periods of improvement or regression as it does in congestive heart failure. (The ascites alone may increase the femoral venous pressure considerably, and the pressure will be reduced somewhat by abdominal paracentesis.

4. Dyspnea may be present to some degree on exertion but orthopnea is strikingly absent when there is no pleural fluid.

5. Cyanosis consistent with venous engorgement is present and may be intense. When combined with lack of orthopnea and marked ascites, the inconsistency of these signs at first suggesting congestive failure is striking. Only marked tricuspid stenosis and pulmonary heart disease simulate it.

6. The heart size is normal in most cases.

7. The heart is free from murmurs except for infrequent apical systolic bruits. The rhythm is usually normal, but auricular fibrillation may occur.

8. The blood pressure and pulse pressure are low.

9. Broadbent's sign is absent.

10. Pleural effusions are common.

11. X-ray pictures of the heart may show calcification of the pericardium, limitation of cardiac pulsation or limitation of pul-
sation of the right border, dilatation of the superior venae cavae, dilatation of the auricles, prominence of the left upper border and pleural thickening. However, the examination may be essentially negative.

12. Electro-cardiography shows low voltage in the axial leads, or inversion of the T waves in leads I and II of the Coronary type.

Burwell and Blalock in considering the diagnosis of Pick's syndrome use the term chronic constrictive pericarditis as applied to the condition. They believe

"Pulmonary edema is rare when pleural effusion is frequently encountered. Unlike most conditions caused by heart failure, these signs remain unchanged for months or years. They point strongly to failure of the right side of the heart and are combined with a normal or only slightly increased area of cardiac dullness, a fixed heart with diminished pulsation, distant heart sound and an absence of visible or palpable apex beat. Usually tachycardia is found and regular rhythm without murmurs. The pulse is paradoxical and the pulse pressure small. The combination of a marked degree of peripheral congestion with a small quiet heart is most weighty in determining the condition."

These men list diagnostic points as follows:

1. The pressure in the veins
   a. The pressure is high (from 150-390 mm water in systemic veins as compared with normal of 50-100).
   b. This elevation is persistent. It may fluctuate but it does not return to normal and it may be high for years.
   c. The pressures in arm and leg are not notably different unless there is considerable ascites, an observation which implies that the obstruction is in the heart and not in the cava.
   d. The pressure rises with exercise more than it does in a normal subject.
   e. The pressure rises with intravenous infusion of saline
7. Edema, ascites, and pleural effusions, which are in the main the result of increased venous pressure. There is no evidence in the analysis of edema fluid, which is poor in protein that the permeability of the capillary wall is increased, nor is the osmotic pressure of the plasma low.

8. Epistaxis — Several of the patients had frequent nosebleeds.

II. Effects of diminished cardiac output and of inability to increase it to a normal extent.

1. Tachycardia, which is in some degree compensatory.

2. Low blood pressure.

3. Low pulse pressure and small pulse.

4. Paradoxical pulse, due partly to the effect of the reduced output per beat (which makes it more obvious) and partly, according to Katz and Gauchet directly to the scar in keeping from the heart the normal effects of changes in intra-thoracic pressure.

5. Weakness and cyanosis. When the output is greatly reduced the patient may feel weak and even exhibit cyanosis and coldness of the extremities, all for an obvious reason. (i.e., reduced flow through the periphery.

6. Lowered tolerance to exercise. This is the most important result of the disturbance of output. The limitation to systolic output deprives the body of one of the methods of increasing the output of the heart per minute. There remains only the method of increasing the pulse rate. This method has rather narrow limits, especially since the pulse is elevated even at rest. Therefore, even mild exercise usually drives the body to an increased removal of oxygen from the blood, and patients who are comfortable at rest
may have severe dyspnea with slight exertion.

White, in a summary of his fifteen cases of Pia's Disease, found the following signs and symptoms:

1. The first symptoms were dyspnea in 4 cases,
2. abdominal enlargements (i.e., a big liver and ascites) in 3 cases,
3. dyspnea and abdominal enlargement in 3 cases,
4. edema of the feet and abdominal enlargement in 2 cases,
5. edema of the feet and ankles in one case,
6. edema of the face and abdominal enlargement in one case, and
7. soreness in right upper quadrant in one case.

In all cases abdominal enlargement was present at some time during the illness; it was the only uniform symptom, but swelling of the feet and dyspnea at some time or the other occurred in most of the cases.

There was little pain or palpitation. Weakness was occasional. Some patients showed loss of weight in the upper part of the body. There was no fever except in 2 cases with subacute tuberculous infections, and in several cases during intercurrent acute infections. In several cases it was clear that the symptoms of acute pericardial involvement passed over into those of chronic constrictive pericarditis.

Two signs were consistently present in all cases:

1. Enlargement of the liver -- non-tender and non-pulsating -- usually with much ascites.
2. Engorgement of the jugular veins with or without evident pulsation. Venous pressure when measured usually was between 20-30 cm. Three times more than normal.
The reason for the preponderance of liver enlargement and ascites in the dropsy of the majority of cases of chronic constrictive pericarditis is probably to be found more in the greater obstruction of the flow in these cases from the hepatic veins at or near their outlet, into the inferior vena cava than of the flow in the vena cava itself. Such obstruction may be due to the pressure of locally constricting adhesions or of fluid (as in acute pericardial effusion), or to anatomical variations, in the venous relationship.

It is to be noted in this connection that dependent or generalized edema antedated or over-shadowed the liver enlargement and ascites in a few of the cases.

The size of the heart was normal in 7 cases, slightly enlarged in 5, and moderately enlarged in 3. In at least some of the cases with apparent slight enlargement, the thickness of the fibrosed pericardium itself could account for the increased measurements by X-ray or physical examination. There were no murmurs in 12 cases; systolic murmurs slight to moderate were heard at the apex in the other three. There were no diastolic murmurs. Heart rhythm was normal in 11 cases, except for some extrasystoles in one; in the other 4 there was auricular fibrillation.

Blood pressure and pulse pressure were low in 11 and low normal in the other 4.

In 7 cases a well marked paradoxical or Griesinger Kussmaul pulse was noted. In 2 cases it was said not to be present; in the other 6 apparently was not looked for.

Broadbent's sign was noted in no cases, nor was inspiratory collapse of the neck veins. (That is a sign of external adhesions).

There was effusion into the pleural cavities in the nature of
a transudate in 7 cases; generally in the more ill cases, and in one other case there was a terminal empyema.

Edema of the legs was present in 10 cases, marked in 4. Abdominal and thoracic walls were edematous in 7 patients, and the face swollen in 2. In no case was the spleen definitely felt.

The heart shadow was normal in size by X-ray in 7 cases and slightly enlarged in the rest. The shape varied from normal to so-called mitral with prominence of the left upper border, and in 3 cases there was indication of left auricular enlargement (two of these had auricular fibrillation). The pulsation varied from fairly free on both sides to little or none anywhere.

It was common to find free pulsation at cardiac apex and left border with no pulsation at the right border which would extend a little too far to the right with a hazy outline.

In most cases there was an increase on supracardiac shadow, apparently due to dilatation of the superior vena cavae and to mediastinal involvement or displacement. In 6 cases there was considerable pleural thickening. Pericardial calcification when noted by X-ray does not mean Pick's Disease. It is a helpful confirmatory sign. The heart was more or less anchored in 6 cases, with or without restricted movements of the diaphragm on one side or the other.

In all 14 cases the EKG record was abnormal in two respects. There was low voltage in 5 or inversion or flattening of the T waves in Leads I or II or both leads in 10 cases. In one case both these anomalies were present. Inversion of the T waves was of the so-called coronary type, like that also found in many cases of acute pericarditis. The EKG abnormalities may persist in whole or part
after operation -- even in cured cases. A complete return to normal appeared in only one case. The reason for the occurrence of these EKG abnormalities is obscure.

The heart rate varied widely. Normal rhythm was the rule but there was auricular flutter in 4 cases.

The blood as a rule showed slight anemia, usually, but not always with a low serum protein. The urine was normal. The liver function was reduced.

Volhard, 1923, quoted by White stated that callous thickening and shrinkage of the pericardial sac result in a typical disease picture with effusion in the abdominal and sometimes in the thoracic cavities. That these effusions at times possess an inflammatory character because of attacks of polyserositis can make more difficult the explanation of the disease pictured, but cannot obscure the predominant influence of the callous pericarditis itself. He said the condition was not recognized because reliance was placed on several signs not pathognomonic. More important to him was the total picture of the disease with its evolution and inflow stasis. He wrote that the most important and conclusive indication was the high grade venous stasis with small heart, overfilling of the neck veins, big liver, and "ascites praecox." He pointed out that in this condition the cardiac difficulty is in the restriction of diastole in contrast to the effect of the important external pericardial adhesions which increase the heart load in systole.

Two doctors give their personal accounts of the symptoms of Pick's Disease, having suffered from the disease themselves.

Whereas Pinee's first symptoms were dizzy spells on exertion, slight
Dyspnea and epigastric tenderness, Finsen's perigastrium and right hypochoondrium upon walking or jumping and fulness in the abdomen following meals. The latter symptom was not relieved by dietary regime or laxatives. Pineo next noted that his collars were tight resulting from swollen veins in the neck. (Froesch also reports this as a symptom in some of his cases).

Later, Pineo experienced swelling of the lower legs and impressions of his belt left on his abdomen. He received Digitalis with no benefit. Pineo's case was reported on through to the cure. One and one half years after the onset a check-up showed the heart normal in size and shape, with the right border hazy and poorly defined without visible pulsation. The pulsation of the left border was feeble. A slight amount of fluid was found at the right lung base. He was then diagnosed as a case of constrictive pericarditis. His pericardium was resected affecting a cure.

Barnes and Harrington believe that the condition is understood readily if one realizes that it results from an inflow stasis of gradually increasing degree. Dyspnea is peculiar in that it is present on exercise but absent when the patient lies flat, and is by no means as pronounced as that observed in the presence of an equal degree of ascites in cases of valvular cardiac disease.

Dr. Blalock studied 10 patients with Pick's Disease and tabulated the incidence of the following symptoms in that group.

- Distended veins 100%
- Enlarged liver 100%
- Tachycardia 100%
- Fixation of heart 100%
- Low pulse pressure 90%
- Increased venous pressure 100%
- Enema (peripheral) 100%
- Diminished pulsations (fluoroscope) 100%
- Paradoxical pulse 90%
- Faint heart sound 90%
Ascites (often early) 80% Pleural effusion 80%
Greatly enlarged heart 0 Pulmonary edema 0
Hypertension 0 Systolic retractions 0
Auricular fibrillations 0 Paroxysmal dyspnea 0

Beck and Griswold⁷⁴ list the clinical manifestations in the order of appearance:

1. Rise in venous pressure (most reliable index).
2. Ascites and development of a fibrinous exudate on the liver.
3. Hydrothorax.
4. Pulmonary and subcutaneous edema. (These are late manifestations).

Blalock⁳⁶ believes that an important sign is the great degree of systemic congestion that was out of proportion to the moderate amount of dyspnea.

Stewart and Heuer⁷⁵ studied 7 patients preoperatively and 5 patients post-operatively. Their measurements showed:

1. The venous pressure was elevated.
2. The circulation time was lengthened (arm to tongue, decholin)
3. The cardiac output (Grollman method) per minute and per beat was decreased.
4. As improvement was observed clinically after operation, there was a fall in the venous pressure shortening of the circulation time, and increase in cardiac output per minute and per beat.
5. Venous distention, observed clinically, was recorded by means of infra-red photographs, and regression of size and number of peripheral venous channels was observed after operation.
Out of 5 operated patients 2 were completely relieved of symptoms and signs, the third was much benefited, but the 2 others who were only recently operated have not shown marked clinical improvement.

Significant diagnostic features found in the above studied patients are enumerated:

1. Restricted or absent motions of the heart (fluoroscope).
2. No sign of enlargement of heart.
3. Calcification of pericardium in 2 patients as shown by X-ray.
4. Paradoxical pulse in all instances.
5. Absence of valve lesions.
6. Normal sinus rhythm in 6 cases and auricular fibrillation in one.

Their conclusions based on this study were:

1. Obstruction to the entrance of blood into the heart shown by piling up of blood on the venous side with increase in venous pressure, and slowing of the velocity of blood flow, since less blood is available to pump, the cardiac output is decreased.

2. The fibrous or calcified pericardium interferes with contraction of the heart muscle (shown by decreased excursions under the fluoroscopic examination); this being the case all the blood which gets into its cavities is not expelled, thus contributing further to decrease in cardiac output per beat and per minute.

Freedman, [76] in discussing diagnosis of Pick's Disease from a radiographic standpoint states:

The experience in the past years demonstrate that the roentgen signs of cardiac compression are more definite than those of pericardial effusion. It is generally recognized now that the roentgenologic examination enables one to exclude many cases suspected.
clinically and to establish a positive diagnosis of cardiac compression in each instance by careful correlation of clinical and roentgenologic signs.

Freedman, from a study of 26 patients reached the following conclusions:

1. Cardiac compression due to pericardial scar formation is usually represented on roentgen examination, by a normal sized or smaller than normal heart. Moderate or marked enlargement of the cardio-pericardial shadow is uncommon and may be due either to an unusually marked thickening of the pericardium or to an underlying valvular or myocardial disease.

2. The majority of the compressed hearts are either triangular or less commonly, globular. In rare instances a compressed heart may show a mitral or aortic configuration.

3. The cardiac pulsations are usually diminished or absent. Occasionally, however, certain sections of the compressed heart may show vigorous pulsation.

4. The aortic knob is flattened or completely obliterated in the majority of compressed hearts.

5. Due to the marked thickening of the pericardium, the heart becomes rigid and in the majority of cases, its configuration remains either unchanged or changes but little during the two phases of respiration.

6. The adhesions between the heart and pericardium lead to a fixation of the heart. The degree of fixation is determined with the aid of expiratory and inspiratory postero-anterior roentgenograms, taken with the patient lying on either side with lateral views taken during both inspiration and expiration.

7. The most conclusive sign of cardiac compression, the cal-

...
cification of pericardium, is present only in the minority of instances. (in 3 among 26 patients).

8. The kymographic examination is of no differential diagnostic value in cardiac compression. It only serves as a record which enables one to compare the difference between the pulsatory amplitude before and after removal of the pericardial scar.

9. Marked change in the configuration of the heart after operation was never found.

Stewart and Heuer in another study of nine patients found the arterio venous oxygen difference before operation was increased in all except one. The range being from 715 cc to 83.6 cc.

After the operation when the patients were in their best state it decreased in all patients and only one fell outside of normal range, which was then 51.4 to 68.7 cc.

The cardiac output per minute and cardiac index (liters per square meter per minute) were decreased in all except one patient, the range of the index being 1.35 to 1.82 liters. After operation it increased and ranged between 1.30 to 2.72 liters and was below normal in only one.

Stroke volume was decreased and the range was from 20-42 cc per beat. After operation it increased and ranged from 33 to 50 cc per beat. The venous pressure was elevated in every case, the range being 17.9 to 24.0 cm. After operation it fell and when the patients were in their best state the range was 8.3 to 16.7 cm.

The arm to tongue circulation time ranged from 13.5 to 29.8 seconds before operation -- in short, it was prolonged.

After operation the range was 7.3 to 17.1 seconds when the patients were in their best state.
There was no consistent behaviour of the heart rate. In certain patients it was elevated before operation and slowed afterwards and in others the reverse happened. The basal metabolic rate was not altered significantly in this syndrome, nor was it changed by operation.

The vital capacity before operation was not lowered if the pleural cavities were free of fluid. In certain patients it decreased and in others it increased after operation. Decrease after operation was in part due to the flexible thoracic cage resulting from removal of ribs. Infra-red photographs revealed marked distention of and increase in the number of caliber of the venous channels before operation. If improvement occurred after operation there was a progressive decrease in their number and caliber.

Their observations showed that chronic constrictive pericarditis is characterized by decreased cardiac output per minute and per beat, rise in venous pressure, slowing of the velocity of blood flow, and engorgement of the venous vascular bed. Fluoroscopic examination showed decreased contraction of the heart chambers, and fixation of the heart was observed. Clinical improvement after operation was associated with changes in all of these functions toward normal. There appeared to be two essential defects in this syndrome, namely,

1. Obstruction to the entrance of blood into the chambers of the heart resulting in decreased filling, and

2. Interference with contraction of the heart. There is evidence for the first in a. the decreased dilatation of the heart in diastole under fluoroscopic examination and at operation, and b. the observation at operation of the thickened pericardium which was not
capable of much distention and may have been calcified. c. Infra-red photographs revealed distention of the peripheral veins. d. The elevated venous pressure is evidence that there is ample blood available for the heart with respect to contraction, impairment is inferred from, 1. decreased extent of contraction on fluoroscopic examination and at operation, as well as from, 2. the examination of the thickened unyielding pericardium incapable of much change during contraction. The two defects result in decrease in cardiac output, per beat and per minute, and piling up of blood on the venous side, accounting for increase in venous pressure and slowing of the velocity of blood flow.

The heart may be unusually small or not much enlarged. (The cardiac silhouette is made up of cardiac shadow plus the shadow of the thickened pericardium).

Removal of the pericardium results in alteration of these two defects, in short, in removing obstruction to blood entering the heart allowing the heart to stretch in diastole and increase in extent of contraction; these account for changes in the circulation which have been recorded after operation.

Stewart and Hauer 73 in another article bring out the following points:

1. In certain cases heart rate was elevated before operation and became slower afterwards, and in others the reverse happened. The basal metabolism rate was not altered significantly from normal in the presence of this syndrome, nor did pericardiectomy change it.

2. The serum protein was low in 2 cases and in the normal range in the others.
3. Distention of the veins of the eyegrounds was marked before operation and decreased with fall in venous pressure after operation. There was limitation of motion of the cardiac chambers and fixation of the heart on fluoroscopic examination.

In considering the symptoms in a group of 9 patients they found the chief symptoms to be swelling of the abdomen, loss of weight, and dyspnea. Two of the cases suffered from an ache in the epigastric region. Distention of the neck veins was present in all, as was enlarged liver. Ascites and edema, although present in the majority of cases was not present in all. Cyanosis was present in the majority and distention of the neck veins was present in all but one. There was noted a paradoxical pulse in all cases. The sinus rhythm was normal in all cases but two where there occurred an auricular flutter. The blood pressure was low in the majority, even normal in 3 cases.

White and Churchill believe that the discussions regarding signs and symptoms of chronic pericarditis are not of much value because there has not been sufficient distinction between the three groups of chronic adhesive pericarditis which they list as:

1. One group where the adhesions are so slight that there is little or no handicap.

2. One group where the adhesions to the chest wall or diaphragm cause cardiac enlargement, cardiac strain and eventual failure.

3. One group where the constricting pericardium obstructs the entrance of blood into the heart with little or no involvement or enlargement of the heart itself.

Churchill believed that a very important sign is the marked
distention of the cervical veins, that do not empty in the upright posture, and especially in the upright position show a very characteristic double collapse in systole and diastole. He quotes Moritz-Tabora as having measured the venous pressure in the arm veins and found very high values of 200-300 mm. of water. This accounts for a marked degree of liver congestion and the tendency to hydrothorax and early ascites, even before edema of the legs supervenes.

Lilienthal believes that since diastole is impeded, the important sign is the never-emptying engorged veins of the neck or even of the arms, and this is particularly noted when the arms are raised above the head. He thinks that it is principally in these cases that the liver swells and ascites is prominent, (the so-called Pick's complex), although Volhard states that the true diagnosis of precordial compression as a cause was never established.

Beck as a diagnostic standard, lists his triad for chronic compression:

1. Small quiet heart.
2. High venous pressure.
3. Ascites and enlargement of the liver.

Beck believes that the compressed heart does not waste any of its energy as do many of the dilated, hypertrophic hearts. He states:

"It functions efficiently, although it is not allowed to function adequately. The cardiac valves in the compression diseases are normal. There are no murmurs.... The efficient, although inadequate action of the heart is an important point in diagnosis. As the compression develops, the systolic-diastolic excursion of the heart is reduced. Sometimes no trace of pulsation can be seen over the precordium.

The compression force upon the heart can never remain at a higher level than the pressure in the venae cavae! Experimentally it was found that the heart could tolerate a higher degree of compression if the compression developed slowly than if it developed acutely. In human patients the compression force may rise to remarkable levels. In my series the greatest
compression force was from 15-45 cm. water.

Why will a compression of 15-20 cm. kill when applied acutely and why can 40-45 cm. be tolerated in the chronic conditions? The explanation lies in the venous pressure levels. There is not sufficient blood in the vascular system to elevate venous pressure above 15-20 cm. in response to acute compression. In chronic condition we have some evidence to show that an increase in the circulating blood volume takes place (dye, etc. in blood). Another possible cause in the elevation of venous pressure is brought about by the accumulation of fluid that slowly forms in the tissues and serous cavities. This fluid compresses the peripheral vascular tree to some extent and elevates venous pressure.

The clinical manifestation of chronic compression of the heart are produced by venous stasis and a reduced arterial circulation. The veins are distended and sometimes exaggerated. They may stand out like goose quills. Cyanosis, ascites, enlargement of the liver and spleen, subcutaneous edema, varicose veins, hemorrhoids, hydrothorax and pulmonary edema are all expressions of venous stasis. The liver becomes cirrhotic in response to long continued circulatory stasis.

Lawrence and Morton present a case, however, in which there is no evidence of venous engorgement even though the venous pressure was increased to 207 mm water. These men believe that the increased venous pressure and the marked increase in arterio-venous oxygen difference in the presence of the classical clinical picture of constrictive pericarditis make the diagnosis of Pott's Disease certain.

Howard thinks that the symptoms of Pott's Disease are chiefly cardiac in origin and consist of dyspnea, cyanosis and dropy. He says that dyspnea is a very varying factor and is usually present only on exertion, except in cases with associated pericardial adhesions and subsequent dilatation of the heart. The most striking objective symptom is often a marked distention of the superficial veins of the neck, chest, and even of the face and upper extremities. Ascites is very apt to occur before the development of a general anasarca, due to a chronic peritonitis from venous congestion, or possibly to an associated hepatic cirrhosis as has been described under the morbid anatomy.
Noth, in regard to the dyspnea found that it was peculiar in that it was a dyspnea of exertion and in spite of the presence of marked edema, the patient is not dyspneic at rest and is not orthopneic.

Hirschfelder advocates an unusual diagnostic aid in Reiss' gastric sounds. In cases where there are adhesions to the diaphragm on listening over the stomach one is able to hear at times the sounds loud and metallic in quality. These sounds are not much influenced by changes of position, by respiration, nor by inflation or filling of the stomach.

Jones and Roberts found reduplication of the second sound at the apex to be a fairly constant finding. Roberts presents a case in which the onset of the disease was sudden and was characterized by repeated vomiting and much flatulence.

Branch and White believe that there is only one sign which is patholgonomic of chronic pericarditis and that is the evidence by roentgen ray of calcification of the pericardium.

McGuire and Havenstein find that the liver was regularly enlarged and firm but usually was not tender and did not pulsate. The heart rhythm is usually regular but irregularity due to auricular fibrillation or extr-systoles is sometimes encountered. A general weakness and low grade fever was occasionally encountered.

Pisula advocates that the feeling of fullness in the epigastrium is due to the stretching of the capsule of Glisson. The anorexia of this disease, he feels, is due to the slowing of the metabolic process because of the poor arterial circulation. The constipation he says, is due to the anoxemia of the intestinal musculature.
Wenckeback\textsuperscript{46} warns us that the pulsus paradoxus is a fallacious symptom because it may be found in perfectly normal people or in other diseases.

McGuire and Havenstein\textsuperscript{34} believe that the reason this condition is so frequently encountered in adolescents and young adult life is probably because unrecognized cases do not survive for many years.

**MECHANICAL DIAGNOSTIC AIDS**

Morton\textsuperscript{35} warns that in order to get satisfactory X-ray plates in these cases one must use three to four times the usual factors.

Beck\textsuperscript{36} believes that when the diagnosis is in doubt exploratory pericardotomy should be regarded as a justifiable procedure. He hesitates to recommend this procedure because of the possibility of inflicting an injury to the coronary vessels with the needle.

Dr. Wagenstein in the discussion of an article by Moses and Barron\textsuperscript{22} informs us that if saline solution is allowed to run from a glass tube into an arm vein with the arm at the level of right atria, the height at which the column of fluid comes to a standstill determines the venous pressure.

Cushing and Feil\textsuperscript{37} in a study of 11 patients found

\begin{itemize}
  \item [a.] Voltage of QRS complex below the usual limits of normal.
  \item [b.] Slurring of the QRS in all leads.
  \item [c.] T Waves of low amplitude -- either of positive or of negative sign. The presence of T waves of normal voltage was an interesting finding.
\end{itemize}

Dieuaide, quoted by Cushing and Feil found that change in position of patients with adhesive pericarditis from one lateral position to the other is not followed by normal change in the electrical axis of the heart.
France, from an analysis of 38 cases in which the shift of the electrical axis was studied during life and the hearts examined at autopsy found that there was no constant relationship between the fixation of the electro-cardiogram and the presence of restricting pericardial adhesions. He came to the conclusion from this study that the electro-cardiographic test for "axis shift" was not of value in the ante mortem diagnosis of adhesive pericardio-mediastinitis.

NORTH found low voltage QRS complex in all leads, the T waves iso-electric in leads I and II and slightly inverted in lead III.

Cushing, in the study of 11 patients found that they all had electro-cardiograms with QRS complexes of low amplitude. The T waves were low in voltage. Following pericardectomy the QRS voltage increased in a number of cases.

Griswold found a small slurred QRS complex. Beck believes that in the presence of Pick's Disease low voltage and slurring of the QRS complex are usually seen in the EKG. Fixation of the electrical axis may or may not be present.

Pilcher and Lond in a study of a case found the only abnormal finding to be inversion of the T waves in lead III.

Stewart and Heuer in a study of nine patients found the QRS complex in leads I to III to be of low amplitude in all but one case in which it was normal. He found that the T wave in leads I to III to be of low amplitude or negative or diphasic. The T wave was also cone-shaped in leads I and II in several cases. The axis deviation was not constant. It was either slightly to the right or to the left.
McGuire and Havenstein found low voltage of the QRS complex. They found the T waves in leads I and II to be frequently of low amplitude or negative. The electrical axis, they believe, may or may not alter with the change of the position of the body.

Dieulaide states that normally there is a change in the form and amplitude of the electro-cardiographic waves in shifting the subject from the right to the left side. This phenomenon is marked in most patients with heart disease. It is due to rotation of the heart. When there is no change in shifting the patient one must think of lesions involving both the pericardium and the mediastinum.

Schwab and Herrmann think that the changes in the RST sector are the result of ischemia of the cardiac muscle. They feel that the inversions of the T waves which follow the changes in the RST segment are associated with organizations and repair in the epicardium and sub-epicardial myocardium.
DIFFERENTIAL DIAGNOSIS

The list of diseases which must be considered before making a diagnosis of chronic constriction of the heart are many. The list includes valvular disease, cirrhosis of the liver, polyserositis, ascites accompanying abdominal new growths, nutritional edema, Beri Beri heart, and large ovarian cyst. Several of these diseases may by complicated by Pick's syndrome and it is here that the test of diagnostic ability arises. The most important factor in a diagnosis of Pick's disease is that a good hope for cure is offered. To opera-tate for compressed heart when mitral stenosis is producing a bizarre syndrome is only to subject the patient to needless surgery and to endanger his life. To treat a case of Pick's disease as if it were a valvular disease is only of value to the druggist who is selling the digitalis. The same holds true for many of the other diseases in the differential diagnosis.

Mitral Stenosis:

Mitral stenosis is the first and most important condition which must be differentiated. This is doubly so because many pediatricians seem to have confused mitral stenosis with adherent pericardium with Pick's Disease.93

McGuire and Havenstein34 in a discussion of the differentiation of these two diseases have come to the conclusion that in mitral and tricuspid disease "enlargement of the liver, edema, and dilatation of the cervical veins are commonly present with tricuspid stenosis or insufficiency, and failure of the right heart secondary to mitral stenosis. The history, the configuration of the heart, and the presence of a pre-systolic or mid or late diastolic murmur are helpful differential findings, particularly as cases of adhesive pericarditis complicated by serious valvular lesions are usually not amenable to surgical treatment.

The picture can be confused with functional tricuspid regur-
gitation which is commonly seen in the acute stages of right heart failure and in chronic right-sided failure secondary to mitral stenosis. The presence of valvular disease, or hypertension, and systolic venous pulsation should serve to differentiate this condition."

Volhard writes in differentiating these two conditions that "a functionally similar or even the same condition occurs in concreto pericardii as in the most severe forms of mitral stenosis. In the latter we likewise find an abnormal output of the left heart, the small pulse of little amplitude, and an intense inflow stasis with the characteristic elevation of venous pressure, liver enlargement and concomitant ascites. The increase in venous pressure corresponds to a marked over filling of the right auricle, which in contrast to the pericardial constriction generally is followed by a permanent irregular pulse. Both the dilatation of the right auricle and the conus pulmonalis have a characteristic roentgenographic appearance. The congested cervical veins not only show a diastolic collapse, but also a systolic swelling, because in this stage of mitral stenosis a functional or organic tricuspid insufficiency regularly exists.... But instead of quietness over the encapsed heart, we feel here the mighty heaving action of the right ventricle over the area of cardiac dullness and under the left costal margin and hear the characteristic mitral rhythm and the diastolic murmur over the apex as it strikes the chest wall. In the presence of these a mistake is hardly possible but it is theoretically of greatest interest that in different ways this combination of an abnormally small cardiac output with stasis on the venous side of the heart produces the same clinical picture and the predominance of portal stasis over peripheral stasis."

Youmans in discussing the different pictures which can be presented by calcification of the pericardium due to various causes comes to the conclusion that "the factor of etiology causes characteristic differences in the clinical picture as well as the pathology. A typical case of tuberculous origin presents the picture of a cirrhosis of the liver with a marked recurring ascites without edema, an enlarged liver and little or no jaundice.

There may be indefinite gastric disturbances, a sense of fullness in the abdomen and occasionally pain, but the patient's general health remains good. The absence of symptoms pointing to the heart is strikingly characteristic in the early stages. Later in the course of the disease there may occur the signs of cardiac failure without evidence of valvular disease. The course of the disease is characteristically slow, insidious and intermittent. A history of tuberculosis is often not obtained."

On the contrary the rheumatic cases present early signs of cardiac embarrassment with evidence of characteristic valve injury. Often there are present the signs of an adherent pericardium. Edema is present from the beginning and is usually marked. A typical rheumatic history is usually obtained."
White is of the opinion that the presence of mitral stenosis may be taken at once as ruling out chronic constrictive pericarditis of any serious grade. Differential points he gives are the diastolic murmur and the enlarged heart and "the very fact that if the heart is enlarged is sufficient to rule out chronic constrictive pericarditis as the fundamental condition". Barron thinks that the marked increase in the venous pressure in spite of cardiac enlargement points against congestive heart failure.

Polyserositis:

The next disease we must differentiate is polyserositis. It has been a common misunderstanding to confuse this with Pick's Disease.

The reason for this confusion is easily understood. Burrell, Hare, et al., in discussing polyserositis believe that "under the generic term 'polyserositis' is included a well-defined group of cases with chronic inflammatory thickening of the membranes lining the great serous sacs, often there are recurrent effusions, and the chief symptoms may be those of congestive heart failure. Many of these cases are regarded as 'probably tuberculous' but clear proof of their exact nature may be difficult to produce. It seems reasonable, therefore, to suppose that chronic tuberculosis of the great serous sacs progressing to the production of 'polyserositis' is a condition associated with a high degree of immunity to the infection, and that in more chronic cases the infective element is slowly submerged as the clinical picture becomes one of mechanical obstruction to the heart and circulation."

White in his discussion tells us, "Polyserositis or an inflammation of several serous cavities, sometimes called Connectos disease is occasionally found in young individuals due to tuberculosis, associated with pneumonia, or most commonly of unknown cause. The pleura and pericardium are the serous cavities generally involved, but sometimes the peritoneum is also affected. The infection begins as an acute process, sometimes insidious and obscure with little clinical evidence and sometimes severe and easily diagnosed; it goes on to a chronic fibrous state of varying importance. Polyserositis is not Pick's Disease although it may be the infection that precedes it in some cases. Neither is Pick's Disease polyserositis although the two frequently coincide. Chronic pleuritis of considerable degree occasionally accompanies chronic constrictive pericarditis as does also chronic peritonitis."
Peri-hepatitis, resulting in frosted or icced liver (Zucker-gaisboeber) first clearly discussed by Gurchohn may or may not be found in Pick's disease; it is not a necessary part of the disease.

If there has been enough peritonitis during the polyserositis that may precede Pick's disease, then the liver may become frosted, or the spleen, or even the intestines.

In cases seen one had this frosted liver. The capsule was smooth and relatively thin in all even though the liver showed considerable nodular fibrosis in one and slightly nodular in two cases.

The most common site apparently for chronic peritonitis is the under surface of the diaphragm. There is a question here as to the possible influence of the blocking of the lymphatic flow in that region to help to explain the persistence of ascites in some cases. Peri-hepatitis, peri-splinitis, and even frosting of the intestine itself may occur quite independently of pericarditis or pleuritis; they may be found associated or as isolated lesions."

Piersol et al.\(^6^5\) came to the conclusion that polyserositis, in a different condition although the two diseases have frequently been confused in the past and may coexist. "Polyserositis, they believe, is associated with a chronic peri-hepatitis in which the capsule of the liver becomes tremendously thickened, giving rise to the so-called sugar-icing appearance. The condition involves little, if at all, the liver parenchyma. The process apparently starts as a low-grade chronic inflammatory reaction of the peritonum about the diaphragm and upper abdomen and gradually involves the pericardium and one or two pleural cavities. The similarity of the two conditions and the fact that mediastino-pericarditis may result from polyserositis makes it obvious that they are not easy to differentiate. Perhaps the two conditions are not essentially different.

On the other hand, from the standpoint of surgical treatment it would seem worth while to try to draw a distinction between the two conditions. In Pick's Disease, if the symptoms are primarily the result of pericardial constriction, cardiolyis would seem to offer more hope of relief than in polyserositis in which the chronic inflammatory process continues even if compression of the heart is temporarily relieved."

Sison\(^9^7\) believes that "Multiple serositis is a clinical syndrome characterized by chronic inflammatory reaction affecting the serous surfaces of the heart, pleura, peritoneum and visceral organs.... Some confuse multiple serositis with the so-called Pick's Disease, but after a careful review of the cases it appears quite plain that in the present state of our knowledge regarding Pick's syndrome, it is entirely a matter of personal opinion and preference what cases should be included in this group of 'multiple serositis'."
Most German and Italian writers are inclined to attribute all the blame to tuberculosis, as the most probably exciting agent in multiple serositis.

The reason these two diseases should be differentiated according to Smith: that the polyserositis is due to an "inflammatory process and of course not at all amenable to the operation of cardio-

Cirrhosis of the liver:

Kelly believes that one can distinguish chronic constrictive pericarditis from cirrhosis of the liver by the absence in most cases of portal congestion and gastric intestinal disturbances — "hemorrhage, diarrhea, hemorrhoids, enlargement of the spleen, marked dilatation of the superficial veins of the abdominal wall..."

Barron tells us that dyspnea on exertion (slightest) is not known in cirrhosis.

White believes that portal cirrhosis of the liver should be easily distinguished from chronic constrictive pericarditis...

"The relative youthfulness of the patient with Pick's Disease, their absence of a history of alcoholism or serous infection or other factor, pointing to cirrhosis of the liver as the chief lesion, their small, often paradoxical, pulse with low pulse pressure, and particularly the apparent conclusive finding of increased pressure in the neck veins quickly distinguishes them from cases of primary cirrhosis of the liver. The changes in the liver in most cases of chronic constrictive pericarditis are relatively slight and consist of vascular congestion plus the so-called cardiac cirrhosis, fibrous strands radiating from the central veins toward the interlobular spaces. The more chronic severe cases show a larger amount of this fibrosis, but not the degree of liver damage seen in primary cir-

Most of the cases have some slight re-
duction of liver function as shown by special tests (such as with Rose Bengal or bromsulphalein) but not the high degree of dye re-
tention found in the presence of jaundice or in a few cases of severe portal cirrhosis of the liver."
Nutritional edema:

This is differentiated from chronic constrictive pericarditis by the increased venous pressure, the preponderant liver enlargement, and the course of the illness. According to White nutritional edema presents quite a different picture "save for the generalized edema, which may occur in some cases of Picket's Disease, and the low serum protein (below 5%) also found in some cases of Picket's Disease, particularly in those with chronic malnutrition. Beri Beri heart:

The generalized edema and signs of right sided heart failure in this condition are theoretically confusing. The history of dietary deficiency peripheral neuritis, tenderness and atrophy of the muscles, the response to Vitamin B is characteristic of Beri Beri. The enlarged liver, fluoroscopic and EKG findings in chronic constrictive pericarditis help to differentiate these diseases."
Disease, especially when chronic, rarely produces a simple change in structure or in function. Physiologic measurements of various sorts that reveal deviation from the normal when applied to patients must therefore be interpreted with caution, for it is not always easy to decide which of the mechanical or chemical changes present is responsible for the abnormal finding.

Burwell and Strayhorn\(^{100}\) found from their studies that the essential defect in Pick's Disease was "the limitation of the diastolic relaxation of the heart by the encircling scar tissue and the consequent fixation of the output per beat at an abnormally low level. The limitation of the output per beat made it impossible for the output of the heart per minute to increase except so far as this could be brought about by increase in the already rapid cardiac rate. The dyspnea the patient experienced on slight exertion was presumably due to the inability of the heart to increase its output adequately and thus resembled the dyspnea suffered by normal persons after severe exertion. The edema, however, was not due to diminished cardiac output, but certainly in the main, to the elevated venous pressure. That this is so is indicated by the distribution of the edema in the patient's body. The volume of blood flow per unit of time was necessarily diminished in both the peripheral and the pulmonary areas, but congestion and edema were observed only in the area drained by the systemic veins in which the pressure is known to have been elevated far above the limits of normal."

These observations are of theoretical importance since they show that the edema is more dependent on increased venous pressure (back-pressure) than on alteration in the volume flow of blood.

Hunter and East\(^{64}\) are of the opinion "that the symptoms of the disease are produced by the mechanical effect on the ventricles; these being constricted cannot fill to their capacity thus producing a back pressure effect on the auricles and a rise in venous pressure, this may affect the inferior vena cava in particular. The effect in the circulation is to slow the rate of the flow of blood between arm and lung."
Beck is of the belief that the heart in Pick's Disease is "a small quiet organ. It cannot undergo dilatation. It cannot undergo hypertrophy.... The anatomical agent producing the compression prohibits the heart from receiving its normal quota of blood for each systole. In as much as it receives a subnormal quota of blood, it actually pumps out a subnormal quota of blood, and the work load of the heart is reduced. The heart can do nothing about this reduction in work and is forced to play a passive role in pumping what blood it receives. I believe that the heart can actually does undergo atrophy of disease in the compression diseases. It undergoes disuse atrophy much as any other muscle undergoes disuse atrophy when its work is reduced."

Burwell and Blalock are doubtful if the heart could expel the normal quantity of blood at each contraction even if it could receive it. They think that the inextensible and relatively unyielding scar, which is attached to the entire surface of the heart must offer a real impediment to systolic contraction. Shortening and rotation of the heart in systole are almost entirely abolished.

Sprague states "The essential physiology of chronic pericarditis in which the heart chambers are fundamentally normal but in which the blood is prevented from entering them either by adhesions about the exits of the venous cavae, or more often by the fact that the chambers are hindered in their capacity to dilate and receive the blood because of the inelastic armour of the thickened pericardium."

Keith in his Hunterian lectures tells us that when adhesions form after pericarditis they begin between the auricles and the pericardium where the great veins pierce to enter the heart and where the structures which close the venous orifices are situated. Keith thinks that "mere adhesion of the auricles to the pericardium, while it may lead to some hypertrophy of the muscular wall, will not markedly affect the normal action of the auricles so long as the outer surface of the pericardium remains free. But when the pericardium becomes adherent to surrounding structures the muscular
bands which surround the venous orifices are greatly handicapped by the adhesions and regurgitation takes place.

The syndrome in Pick's Disease and chronic intracardial pressure can be produced by:

1. Scar over entire heart.
2. Scar over ventricles.
3. Fluid in pericardium.
4. Fluid inside, scar outside.
5. Scar glazed over heart, fluid outside and scar outside fluid.
6. Band of scar over the ventricles.
7. Band of scar over auricles.

The scar tissue which is laid down on the epicardium or parietal pericardium in subject to subsequent contracture. When shrinkage takes place the heart becomes compressed. Studies by Cutler seem to indicate that the accumulation of fluid in either the thorax or abdomen is dependent upon whether the right or left heart is impeded.

Burwell and Flickinger, as a result of studies regarding cardiac output and the arterial venous difference of oxygen in cases with constricting pericarditis, came to the conclusion that "such diminution in total cardiac output may be expected to lead to weakness and fatigability. The small output per beat cannot be increased in this condition, and this limitation may be expected to lead to tachycardia, a low pulse pressure and a limited tolerance for exercise. The diminution in cardiac output is thus important factor in production of symptoms."

These men also believed that the increase in venous pressure was
an important factor and a sufficient increase in this pressure could lead to venous distention, to engorgement of the liver, to the formation of peripheral edema and to the transudation of fluid into the peritoneal and pleural cavities.

Burwell and Blalock 71 believe that the elevation of the venous pressure and the decrease in cardiac output do not bear a simple relationship to one another, because the elevation of the venous pressure presumably increases the diastolic filling of the heart and thus tends to maintain or elevate the cardiac output per beat. Conversely, they think there is some evidence that lowering of the venous pressure (i.e., by diuresis or phlebotomy) may reduce the cardiac output.

These men have evidence that the high venous pressure is more important than the low cardiac output in producing disability.

Cranfield, Gwyn, et al 104 state that the heart remains small as a result of the constricting pericarditis and its activity is more impaired in diastole. The inflow stasis, they have found, is produced by two causes: first, constriction about the orifices of the great veins, and second, restriction of diastolic expansion of auricular and ventricular chambers. The reason the heart does not hypertrophy, according to them, is because of the interference with the coronary circulation. Mott 40 also believes that the reason the heart does not undergo hypertrophy is because of the interference with the coronary circulation. McGuire and Havenstein 34 are of the same opinion stating "It is likely that encasing mass of fibrous tissue at times interferes with blood flow through the coronary arteries and may in part account for fibrosis and degenerative changes in the myocardium usually found in this disorder."
Adhesions:

The factor of adhesions -- whether or not they are of an influence in this syndrome -- in fact, whether or not they cause the syndrome -- must be considered. This factor perhaps more than any other is responsible for the lack of understanding and agreement regarding Pcks' Disease.

Beck,34 from an experimental and clinical basis, believes that "it can also be said that extra-pericardial adhesions are silent, and that while both of these lesions combined can increase the work load of the heart, still these combined lesions usually, but not always, are silent. A second way in which extra-pericardial and intrapericardial adhesions combined can affect the circulation is by angulating the heart from its normal axis. A third possibility is by the development of cardiac compression, but in this condition the compressing scar is the detrimental factor while adhesions, when present, are entirely silent and incidental. In general it can be stated that the importance of cardiac adhesions has been greatly over-emphasized in the past. In most cases the produce no circulatory embarrassment whatsoever."

McGuire and Havenstein,34 in a study of autopsy material at the Cincinnati General Hospital for a period of 13 years, found

1. Adhesions binding the visceral and perietal pericardium firmly together either when unassociated with external pericardial adhesions, or when associated with adhesions firmly binding the heart to the diaphragm, lungs, or mediastinum only are associated with cardiac hypertrophy when complicating hypertension or some valvular lesion co-exists. This factor can explain the large hearts sometimes found with adherent pericardium.

2. External adhesions alone extending from the pericardium to various intra-thoracic structures do not cause and are not associated with cardiac enlargement.

Ochsner and Herrmann36 believe that when external adhesions are present, internal adhesions are always associated and that it
it is possible to have complete synechia of the pericardial sac without any evidence of external adhesions.

Simon in an attempt to explain the factors regarding adhesions submits the following reasoning:

"The vena cavae when it leaves the diaphragm, is extra-pericardial and is covered by fibrous tissue... It may be (fibrous tissue) a derivative from the diaphragm or from the fibrous bag of the pericardium or independent of both. It is not covered by the pericardial serous sac until about one half inch before it enters the right auricle. This three fourths inch of vena cavae which has as yet no connection with the serous sac of the pericardium, is therefore very likely to be affected by a proliferation of connective tissue, such as occurs in mediastinitis; but it is equally obvious that adhesions of the two layers of the serous sac, -- namely, that overlying the heart muscle and that lining the fibrous bag of the pericardium -- cannot affect this part at all.

It is possible therefore, to understand why internal adhesions of the pericardium may occur without inducing any signs of pressure on the cava, such as obstruction to the entrance of the blood into the right auricle, and consequent increase in size of the liver. When such signs occur it would follow that we may assume that external adhesions have been formed. This interpretation, if correct, would explain why sometimes chronic pericarditis may exist for years without producing any symptoms until gradually those of myocardial degeneration assert themselves, and, on the other hand, why very rapid signs of heart failure without general valvular lesions may probably be found to be dependent on external pericardial adhesions."

Broadbent writing in 1895 presents the modern view of the group who advocate the effect of adhesions in producing symptoms. He says,

"The heart itself may be enlarged and hypertrophied or atrophied, or normal in size... When the heart is apparently atrophied, the pericardial adhesions are usually dense and firm, and appear to have, so to speak, strangled and compressed the heart so as effectually to prevent enlargement of it, or even its normal development, in the case of a child who arrives at maturity. It is more difficult to account for the enlargement of the heart that is often found in association with, and is apparently due to, adherent pericardium....

Why, then, is the heart normal in size in some cases, and considerably enlarged in other cases of adherent pericardium? The explanation I would suggest is the following: When the heart is found to be dilated and hypertrophied, there being no valvular disease to account for it, it is due to the fact that it has been left in a condition of dilatation after the original attack of pericarditis and that while in this condition of dilatation the pericardium has become adherent; then the adhesions becoming organized, the heart
is effectually prevented from again recovering its normal size. Subsequently it undergoes some hypertrophy.

The following are the steps in the process: During the attack of pericarditis the heart becomes considerably dilated in consequence of the myocarditis accompanying it; after the subsidence of the attack the heart remains enlarged. It is obvious that the fact of the heart being enlarged and dilated would favor the formation of adhesions by the approximation of the walls of the heart and the pericardium. A further contributory cause will be the fact that the heart, in a condition of dilatation, beats with less force and has less power to free itself from adhesions when they form. Again, as the cardiac dilatation is due to myocarditis accompanying the pericarditis, the contractile power of the heart must be impaired, not only at the period of the attack, but for some time after, so that it remains dilated until the pericardial adhesions have become organized into firm unyielding fibrous tissue, which prevents it again contracting down to its normal size.

This would explain why the heart should in some cases of adherent pericardium be found considerably dilated and hypertrophied. The explanation why, in other cases, the heart should be of normal size would be that it had not dilated during the original attack of pericarditis, or else had recovered from its dilatation before adhesions were formed."

Fenton and Vacuez, writing at a much later date, present views almost identical with those of Broadbent.

Alburt, a present day authority, writes, "In these large, practically complete adhesions the state of the heart is not constant; mechanical causes combine variably with the pathological. The prevailing event is hypertrophy and dilatation in various relative proportions. In some cases we are not surprised to find an atrophy. Much depends of course on the original infection and the age of the patient; disease of the endocardium, the patency of arterial channels, and so on.

Some time, however, may elapse before any symptoms attributable to pericardial adhesions may appear...

It would seem that valvular defect is apt to arise in these cases of synochin, as a so-called "functional" derangement, a derangement due to distortion of the heart rather than to static alterations in the valvular apparatus itself which may or may not be present."

Younes believes that adhesions differ in the tuberculous and rheumatic forms of adhesive pericarditis. According to him, the tuberculous type of adhesion are rarely attached to the chest wall and evidence of the involvement of the pleura and peritoneum are common. "Indeed the peritonitis may be the cause of the presenting symptoms.
Not infrequently the pleural, occasionally the peritoneal involvement is inactive. A striking finding is the rather frequent occurrence of a true atrophic cirrhosis of the liver, which may explain in part the ascites. It is suggested that this is due to actual involvement of the liver by the disease process, is not secondary to heart failure, and that it offers evidence in opposition to the idea that the cardiac embarrassment is the principal factor in the causation of the ascites and liver changes. Perisplenitis and sometimes peri-hepatitis is encountered. In the rheumatic type, adhesions of the pericardium to the chest wall and diaphragm are common. The heart shows typical rheumatic lesions and the changes in the abdomen are the result of heart failure.

Smith and Liggett have exactly the opposite idea. They state, "Adhesions of rheumatic origin are more likely to be intra-pericardial while those of tuberculous etiology, are generally extra-pericardial."

Calcification:

We have seen that when calcification of the pericardium can be demonstrated by X-ray in the presence of certain signs and symptoms it is pathognomonic of Pick's Disease. Yet calcification of the pericardium in itself does not mean Pick's Disease. In the majority of cases there is a paucity of clinical findings. The age group is different from Pick's Disease in that 72% are over 40 years of age. Pre-cardial pain is present in about 42% and is due to direct involvement of the cardiac nerve by the calcified pericardium. This is a rare sign in Pick's Disease. There were only 35 cases of calcified pericardium
diagnosed during life up to 1932 as compared with a much larger number diagnosed as Picks' Disease.

The etiological factor when studied, however, shows a close relationship to Picks' Disease. There is strong evidence in favor of tuberculosis in the majority of the cases, and although cases due to rheumatic virus undoubtedly occur, they are less common and possibly the streptococcus or some other organism such as the pneumococcus is occasionally responsible. According to Youmans it seems probable that the majority of cases of calcification of the pericardium represent a variation in the course of Picks' Disease due to some, as yet, unknown factor.

Smith and Willis believe that calcification of the pericardium is a sequel of extensive chronic adhesive pericarditis and is an end result of the same inflammatory process that produces chronic adhesive pericarditis.

In about 10% of the cases of adherent pericardium, calcification is present. This calcium has the same composition as when found elsewhere in the body.

Out of this 10% we may assume with Youmans, who in a study of 104 cases found "that there exists, however, a group of cases in which there is a definite and well marked calcification of the pericardium, associated with quite constant changes in other tissues. During life these cases exhibit a characteristic clinical syndrome, which, with the pathologic changes, warrant their inclusion in a definite disease group. To summarize the findings briefly these cases show pathologically an oblitative pericarditis with calcification frequently so marked as to deserve, the term 'armored heart'. In addition, there is usually found chronic inflammation of the pleura or peritoneum or both, and all grades of changes in the liver, from single chronic passive congestion to true atrophic cirrhosis, including perihepatitis and occasional perihepatitis. The heart, as a rule, shows either no abnormal findings or else degenerative changes in the muscle itself. Clinically these cases exhibit characteristically, in the early stages, the picture of cirrhosis of the liver; marked and frequently recurring ascites without edema of the legs, enlarged liver with little or no jaundice, a sense of fullness in the upper abdomen, digestive disturbances, and occasionally pain.
To these may be added, although they usually appear late in the course of the disease, the signs and symptoms of cardiac failure, as a rule without any evidence of any particular valvular disease. The course of the disease is characteristically slow, insidious and intermittent.

Stokes believes that atrophy is an invariable accompaniment of calcified pericardium and due to mechanical causes.

The similarity of the cases of calcified pericardium to that group of cases called Pick's Disease is even more striking when one considers that in many of the reputed cases of Pick's Disease, are in addition cases of calcified pericardium, notably so in two of Pick's own cases. One is, therefore, justified in stating that when calcification of the pericardium produces symptoms, they are those of Pick's syndrome -- that although the age group is more advanced in cases of calcified pericardium, the symptomatic group occur at the younger age levels, and that calcification of the pericardium represents an advanced stage of Pick's Disease probably associated with a primary polyserositis.

Respiratory system:

The respiratory findings in Pick's Disease depend on whether or not the left ventricle is involved. If it is the disturbance is most felt "in the lesser or pulmonary circuit in the way of dyspnea, asthmatic symptoms and pulmonary edema...." 

Beck, arguing from experiments made by his assistant Miss Maltby, where it was found that the amount of blood put out by the heart could be reduced to 50% and the actual output in the adult per beat could be as little as 25 cc, feels that "the dearth of oxygen in the sluggish venous circulation is a stimulus to the respiratory center and dyspnea may be produced."
Keith, in an outstanding article on the physiology of the heart in various diseases observed:

"When the pericardium becomes fixed by adhesions to the lungs and chest wall a number of obscure symptoms follow owing to the arrest of the normal respiratory movements of the heart. The type of respiration may change; the patient instead of gaining his inspiratory space by a forward heave of his abdominal wall, due to the descent of the diaphragm, inspires by expanding the walls of his chest and body in a lateral direction. Further, in some of these cases, especially if the inflammatory process and adhesions have been confined to the structures of the posterior mediastinum between the base of the heart and the spinal column a peculiar type of heart action -- pulsus-paradoicus -- may appear. These conditions find their explanation in the distended respiratory movements of the heart."

Paradoxical pulse:

The Kussmaul or paradoxical pulse is an interesting finding in the syndrome found in Pick's Disease. Kussmaul in 1874, in his original description, made the following observation:

"Clinically our affection of chronic inflammation of the pericardium and its obliteration, which is a criterion of mediastinitis, leads to a peculiar pulse phenomenon from time to time associated with unusual behaviour of the neck veins. During the time that the sternum with each inspiration exerts a narrowing tug upon the ascending aorta or the arch, the pulse in all the arteries becomes regularly and rhythmically smaller, while the heart movements remain constant. Thus with each inspiration at regularly repeated intervals, the pulse becomes smaller to return again with expiration. I propose, therefore, to call this the paradoxical pulse, partly because of the peculiar disproportion between the heart activity and the arterial pulse; partly because the pulse, in spite of seeming irregularity, in reality has become a regularly recurring waxing and waning.

Kussmaul felt that the fibrinous growth tended to produce in the mediastinum "a callous skein and compact threads, which extend upward from the pericardium to the arch of the aorta, innominate vein, surrounding the stem of these vessels, drawing, kinking and twisting them, binding the arch against the pericardium, and joining the vessels directly to the upper part of the breast bone. The pulse in all of the arteries will be small or disappear entirely
during inspiration, and return to normal during expiration." An interesting observation made is that with this phenomenon the veins in the neck, especially the bulb of the jugular vein, will be swollen during inspiration.

Hirschfelder, writing at a later date substantiates Kussmaul's theory by experiments on dogs.

Schreiber, in distinguishing the true from the false Pulsus paradoxus, gives these criteria:

1. It must be felt in all accessible arteries.
2. It does not require a deep inspiration.
3. There must be no irregularity of the heart action.

Gauchat and Katz considered the occurrence of this phenomenon and the abnormality of the respiratory tract and believe it to be produced by modifications of the intra-pleural pressure. The experimentally showed that adhesions need not be attached to any particular vessel, but may be fastened anywhere between cardiac structures, and thoracic parietes, and still be capable of producing pulsus paradoxus. They also showed that mechanical compression of the aorta produced an immediate effect upon the amplitude of the pulse. Compression of the pulmonary vein caused a decrease in pulse amplitude after one or two beats and compression of the vena cavae had the same effect after three or four beats. They believe that these time relations show that the pulsus paradoxus is always the direct or indirect result of parietal occlusion of the pulmonary vessels, the aorta, or both, and never due to compression of the vena cavae. They think that a paradoxical pulse probably appears in both the pulmonary and systemic circuits, and that the
arterial pulsus paradoxus is due to the impaired flow into the left ventricle.

Ascites without edema:

The presence of the marked ascites with little or no extremital edema, is a point of great interest. Rolleston explains the mechanism of the ascites as follows:

"The pericardial adhesions by contracting, lead to dilatation of the right auricle, inferior vena cavae and hepatic veins, and by this means free regurgitation of blood into the liver is rendered permanent. It is possible that at the time of the primary pericarditis inflammation spreads to the mouth of the hepatic veins and by weakening their walls leads to dilatation and so to a freer entry of blood into them. When once brought about, this dilatation of the hepatic veins becomes permanent. The brunt of the backward pressure thus falls on the liver, while the other branches of the inferior vena cavae, the renal and iliac veins, suffer less than in ordinary cases of chronic engorgement of cardiac origin." 117

Eisenmenger quoted by Wells 54 attributes the ascites without edema of the legs to conditions outside the liver, chiefly, torsion, compression or angulation of the inferior vena cavae by the pericardio-mediastinal adhesions; or else through a localized peritonitis, with some possible etiological relation to the pericarditis, at the transverse fissure of the liver.

Roberts 118 has pointed out that in more typical cases when the disease does not extend beyond the pericardium some further explanation of the predominance of the ascites over other signs of obstruction to venous return must be sought for. He suggests that the ascites predominates, since the liver is entirely dependent on the blood being sucked out of the liver when the chest expands, whereas all other parts of the body have some additional means of getting blood toward the heart. To quote:

"The accepted explanations, I think, do not suffice because they depend on anatomical fallacies. It is supposed that the
hepatic veins are more compressed than is any other part of the vena cava, but there are no hepatic veins outside the liver, and so they cannot be compressed in the pericardium."

Simon believes the limitation of diaphragmatic movement has a very important bearing on the liver. Physiologically it is compressed by the descent of the diaphragm, and in its turn presses on the vena cava, so that the blood from the hepatic veins is pumped up through the inferior vena cavae into the right auricle. If the diaphragm does not act, this free passage of blood does not take place; it is dammed up in the liver which becomes swollen.

Wenkebach has given a quite plausible explanation of the presence of ascites without edema of lower extremities which is as follows: He believes it possible that abnormal inspiratory traction upon the crura of the diaphragm compresses the blood in the upper part of the inferior vena cavae and thus leads to undue distention of the hepatic veins and ultimately to hepatic obstruction and ascites (especially as there are no valves in the hepatic veins).

Weiss, quoted by Kelly, attributed the cause of the disproportionate ascites to changes in the blood vessels of the peritoneum, the result of chronic peritonitis. He believed that in consequence of the pericardial obliteration, general venous congestion occurs; that in consequence of the inflammation of the peritoneum, the peritoneum becomes a punctum minimae resistentiae; that transudation of serum occurs more readily from the altered vessels of the peritoneum than from other vessels of the body; and that in consequence of the development of the ascites venous congestion in other parts of the body is relieved, and the development of edema more or less presented.
Harris, also quoted by Kelly, believes that the explanation of the occurrence of the ascites is that a chronic peritonitis ensues and to it the ascites is due. He feels that such chronic peritonitis in some instances may possibly be an independent affection, and one not directly connected with the mediastinal or pericardial lesion; in other cases it is conceivable that the chronic venous congestion due to the intra-thoracic effects sets up the chronic peritonitis.

Smith and Liggett, in a study of 107 cases found that the incidence of the presence of ascites without edema of the lower extremities was 54%.

Beck found that when the venous pressure reached 16 cm. of physiological solution of NaCl the liver becomes palpable and ascites appears.

Ramsey tells us that the ascites rapidly appears after tapping. The ascitic fluid, he says, is usually of a yellow or amber tinite showing albumin. The specific gravity is 1.015 or higher.

Liver:

Rolleston believes that the liver in Pick's Disease shows advanced changes of chronic venous engorgement with fibrous replacement especially under the capsule, where to the naked eye the appearance suggests chronic thickening of the peritoneum. The difference can, however, be seen at once by microscopic examination.

Elliott and Nadler are of the opinion that typical Pick's Disease does not show a peri-hepatitis. The apparent capsular thickening, they think, is due to a great increase of fibrous thickening beneath the capsule. "When actual capsular thickening
is present this is believed to be accidental; such cases are con-
considered as occupying a position midway between pseudo-cirrhosis and
multiple serositis.... Fibrosis is confined to a small area under
the capsule where, atrophic liver cells are replaced by scar tissue
and to small scattered islands where the existing, connective tissue
appears prominent because of atrophy of hepatic cells."

Sprague et al. 38 believe that the early and preponderant liver
involvement in constrictive pericarditis is due not to a peri-
hepatitis which may or may not be present (depending on the oc-
currence or absence of polyserositis) but to the greater tendency
for obstruction to blood flow out of the hepatic veins than in the
inferior vena cavae itself. The reason, they think, for this ten-
dency for obstruction to blood flow out of the hepatic veins is be-
cause

1. Pressure in hepatic veins less than in inferior vena cavae.

2. The hepatic veins empty into the inferior vena cavae just
below the diaphragm at an acute angle which is made easily more acute
with resulting narrowing of the mouths when there is pressure from
above by pericardial adhesions or fluid.

Volhard, quoted by Rothstein, 25 i.e. of the opinion that the "hep-
atic congestion, enlargement, and eventual cirrhosis are generally
considered as due to hepatic vein obstruction resulting from the
kinking and the compression secondary to the chronic mediastino
pericarditic adhesions obstructing the large veins of the caval
system.

Willius 63 found that there was no interference with the function
of the liver from obstruction except for its circulation.
The type of cirrhosis one finds in the liver in this condition is the so-called cardiac cirrhosis -- that is, "the only type of cirrhosis in which the increased fibrous tissue is found at the center rather than at the periphery of the lobules." 120

Semenov, quoted by Keith,121 thinks that the liver tenderness which is found, at times, is due purely to venous back pressure.

The respiratory mechanism is closely connected with the size of the liver since with improvement of this mechanism there is a decrease in the size of the liver.46

Polyserositis:

Kelly 29 who has written a masterpiece on multiple serositis is quoted directly on his ideas regarding this condition:

"Now, what I believe is that what is true as regards experimental work and acute infections is true also as regards chronic infections; that is, the attempt on the part of the peritoneum to remove certain noxious agents -- for instance, the tubercle bacillus, the infective agent of rheumatism, and other diseases, probably certain toxic substances -- may result in partial or complete success. In the latter instance the peritoneum may be completely rid of the infective agent, which, being carried to the mediastinal lymph vessels and lymph glands, may infect the pericardium or the pleura, giving rise to a primary pleuritis or pericarditis. Subsequently the peritoneum may become infected. In other cases the attempt on the part of the peritoneum to remove the infective agent being only partially successful, the region about the liver and the under surface of the diaphragm suceeds and a primary peri-hepatitis occurs. Subsequently the infective agent may travel through the diaphragm and infect the pericardium, or the pleura or both. In these cases, on account of the peculiar lymphatic supply of the surface of the liver and the under surface of the diaphragm the infective agent being as it were concentrated to the region about the liver, especially to the neighborhood of the suspensory ligament toward which many of the lymphatics converge, gives rise to the excessive and often hyperplastic lesions sometimes observed."...

I believe that in the majority of the cases primary importance in the production of ascites cannot be assigned to the liver. Importance of perihepatitis and the peritonitis in causing the ascites and that in consequence of the inflammation, the peritoneum becomes a locus minoris resistentiae; that in consequence of the chronic pericarditis -- in reality the pancearditis -- the functional activity of the myocardium is interfered with; that in consequence of the
Chronic pericarditis—in reality the pancarditis—the functional activity of the myocardium is interfered with; that in consequence of the lessening of the functional activity of the myocardium the circulation of the blood is more or less impeded—in some cases insensibly impeded; that the region that first manifests the sensible evidences of impeded circulation is the locus minoris resistentiae—the peritoneum; that the relief afforded the general venous circulation by the accumulation of the ascites, probably indirectly, prevents to some extent the occurrence of oedema of the extremities, that the connective tissue hyperplasia of the liver in some of the cases is due to the same causes that induce the serous membrane inflammation—the irritant being carried by the lymphatics into the liver substance from the diseased capsule; and that in different cases the ascites is increased by the contraction of the nearly formed connective tissue of the capsule, by the concomitant peritonitis in the transverse fissure of the liver should it compress the vessels (which occurs rarely if at all), by the marked congestion and its consequences, by concurrent cirrhosis (which is unusual), and by the general failure of the circulation toward the close of life.

De Renzi quoted by Ramsey in discussing the polyserositis believed the peritoneum to be usually involved first, then the right pleura and then the pericardium, and if the right pleura was involved first then that the disease extended to the peritoneum and thence to left pleura and the pericardium. Whether the lesion begins above or below the diaphragm the ultimate lesions are generally the same and the condition is essentially a chronic multiple serositis, which he thinks, generally pursues a remarkably slow and insidious course giving rise to exudation of large quantities of sero-fibrinous fluid.

"The fluid portion of the exudate is generally reabsorbed, leaving the fibrinous substance deposited upon the involved viscera. This fibrinous deposit is then invaded by leucocytes, mast cells, and fibro blasts; blood vessels are scanty and there is a tendency toward lamellar formation.

This peculiar over-growth, of fibrous tissue may become hyalinized, giving the deposit a glistening appearance; it may even resemble a cartilagenous substance and the deposit of lime salts may give rise to the formation of bone like plaques.

Dense adhesions to the surrounding tissue may undergo cicatrical contraction."
Beck, however, is of the opinion that when the organs become covered by a layer of fibrin and fibrous tissue that this condition is not produced by infection, but that venous stasis without infection can produce it. In his experiments he showed that a perihepatitis usually follows, "a chemical irritation of the pericardium and that ascites is a consequence." This fact he feels demonstrates that the pericardium and Glisson's Capsule are in close relationship, and what "affects one may affect the other."

Experimental Work:

Hosler and Williams experimented on 26 dogs by attaching the pericardium to the diaphragm and the ventricles to the diaphragm and then vigorously exercising the dogs on treadmills. They also studied 47 autopsy cases with extensive pericardial adhesions. From these studies they came to the conclusion that "adhesions to the chest wall do not cause hypertrophy contrary to the common accepted belief— and as paradoxical as it may seem, we are led to believe that adhesions per se do not cause circulatory embarrassment, unless they are extensive enough to cause cardiac compression. We believe they have been overemphasized in the past."

Blalock and Burwell produced Pick's disease in two dogs by the introduction of aleuronat into the pericardial cavity. They found that the thickening and fusion of the pericardium were associated with a dilatation of the thoracic duct, with a lymph pressure elevated to approximately the same extent as the venous pressure, and with a small amount of blood in the proximal end of the duct. They drew no conclusions as to the part the elevated lymph pressure plays in the accumulation of fluid in the serous cavities.
These men also found the cerebrospinal fluid pressure greatly elevated.

Beck and Griswold\textsuperscript{74} produced fully developed Pick syndromes in dogs without any adhesions between pericardium and epicardium. In these cases the impediment to the circulation was found to be produced by scar tissue formation in the parietal pericardium, without any adhesions to the heart. Their conclusions were that:

"The essential factor in the production of Pick's disease, as can be seen from the experiments, is fibrosis and contraction of the parietal pericardium or epicardium or both, forming a casing of scar which compresses the heart and primarily obstructs its filling, interfering with cardiac motion as a tightly fitting glove impairs any free movement of the hand. Generalized adhesions between the epicardium and the epicardium and the parietal pericardium are not sufficient in themselves to produce polyserositis. Such adhesions can be produced experimentally, and similar conditions are found clinically in which polyserositis does not develop. Nor is generalized adhesions necessary for the development of polyserositis. A marked degree of polyserositis developed in one experiment in which there were no adhesions to the heart."

Walby\textsuperscript{125} from experiments on the cardiac output introduces the term cardiac index, by which is meant the cardiac output per square meter of body surface. The normal, according to her, is $2.2 \pm 0.3$ liters per minute. Any deviation greater than 15\% when determined by the acetylene method she feels is pathological and is strong confirmatory evidence of constriction of the heart.
TREATMENT

Treatment of the compressed heart consists essentially in surgery. Pisula\textsuperscript{84} expresses this well when he says "a dilated heart is a medical disorder. A compressed heart is a surgical disorder."

In the literature one finds many instances of this condition being treated by medical means such as digitalis; diuretics, high protein diet, restriction of fluids, etc.

K. Tice,\textsuperscript{43} Elliott and Nadler\textsuperscript{119} advocate the use of digitalis. Stewart, Heuer, Dietrick\textsuperscript{77} et al. however say that the "use of digitalis appears to be contra indicated except under certain circumstances."

These men state that digitalis decreases cardiac size and increases ventricular contraction. In the presence of chronic constrictive pericarditis the size of the heart may already be restricted and the cavities small and further decrease in size may not be beneficial, but may increase the obstruction. On the other hand the heart is probably contracting as fully as possible while it is attached to the unyielding pericardium.

Lawrence and Morton\textsuperscript{80} also state that Digitalis produces no beneficial effect on patients with this disorder.

Churchill\textsuperscript{22} in discussing this problem states "If improvement results from digitalis therapy it is probable that the myocardial insufficiency is playing an important role in the case in question. If improvement does not follow, rest and administration of digitalis the mechanical factors may be considered paramount; but an underlying myocardial weakness cannot be excluded."

Stewart and Heuer\textsuperscript{73} warn us that "Venesection appears to be contra
indicated in the presence of this syndrome, since the elevated venous pressure may represent the head of pressure which is required to maintain the circulation."

Stewart, Heuer, et al.77 Preoperatively keep the fluid intake down to 1200 cc, the salt to 2 grams a day, and a high protein diet.

Picard84 advises that any drug which depresses the respiratory center is contra indicated in cardiac compression.

In planning the operation of decortication of the heart, certain physiologic considerations have more than a mere theoretical bearing on the question. The unfortunate results in a number of the cases of pericardial resection that have found their way into the literature have emphasized the importance of having clearly in mind an accurate knowledge of the pathologic changes in the individual cases.

One is forced to admit at the outset, that no reliable criteria exists for determining the capability of the cardiac musculature to withstand the suddenly increased load. Numerous observations and experiments have demonstrated that under the conditions of normal life the pericardium plays no important part in supporting the wall of the heart. Conditions may be different, however, when the myocardium has been damaged by infection. Further, in an individual case it may be difficult to differentiate the symptoms of cardiac failure due to mechanical factors from those referable to myocardial weakness.22

Two factors which must be taken into consideration are rheumatism and tuberculosis. We have seen that "if rheumatism can cause Pake's disease it does so only in the rarest cases."10 This fact is of utmost importance to the surgeon, "and should lead him to question
seriously the propriety of undertaking the operation in patients with rheumatic heart disease. While rheumatic fever not infrequently causes obliteration of the pericardial cavity, the evidence that these adhesions may produce the constrictive pericarditis syndrome is slight indeed. Active tuberculosis of the pericardium may produce the entire syndrome of chronic constrictive pericarditis. The point at issue is whether operation can be effective if performed during the active phase of the infection."

Regarding this last point the reports in the literature of operations performed during this period are uniformly discouraging. Blalock in a study of 43 patients strongly suspected of having or proved to have tuberculosis of the pericardium found, "Most of the patients were over 40 years of age. A positive diagnosis was made as a result of aspiration operation, or autopsy on 25 patients. The pericardium on 2 of these patients was drained during the acute stage, under the mistaken impression that there was a pyogenic infection and these patients died a number of weeks later. The only patients in this group who are living are 4 of those on whom pericardiectomy was performed after the fluid disappeared and a more chronic stage of the disease was reached. However some activity was still present. This is not meant to imply that all patients with tuberculosis pericarditis die unless operation is performed. Tuberculosis pericarditis was strongly suspected but not proved for the remaining 18 patients, who were in general less ill than the others, 8 of these are dead, 4 are free of symptoms, and the remaining 6 are wholly or partially incapacitated."

The diagnostic problem is, therefore, to recognize the active phase of this disease and postpone operative interference with the belief that the best chance for life lies in a self termination of the active tuberculosis process. "If and when activity subsides and the patient is left with a healed scar constricting the heart, surgery can be successfully undertaken."

Of equal importance to the state of the myocardium is the matter of balance between the actions of the right and left ventricles. Churchill believes, "It is of the greatest importance to ascertain the extent to which the scar involves these two chambers. If the output of the right ventricle is suddenly increased by decortication while the left
ventricle remains in scar, the area of venous stasis is merely transferred from the caval system to the lungs and disaster follows."

Fluoroscopic examination in different positions with special observations of the ventricles is invaluable in ascertaining the extent to which each is involved in the scar. Also information will be gained regarding tubercular effusions.

Schmieden in discussing this problem states that the, "Degree and extent of scar formation and disturbances in the coordination of various parts of the heart can be estimated to a certain degree before operation. When the heart action on the left side is just slightly impaired there is nothing more than damping back of the inflow on the right side, as stated by Volhard. Careful roentgenological observation of the heart wall show the heart contour absolutely rigid and motionless on the right and the superior vena cavae as a broadened band due to back flow.

The left side still shows good motion. If the incarceration of the left heart has progressed to a greater extent, clinical symptoms of a left-sided inhibition of inflow will prevail. Great air hunger, oedema of the lung, and bilateral hydro thorax, which can hardly be checked by pleural puncture. In such cases the X-ray will show weak excursions of the left heart also."

Schmieden, one of the pioneers in this field who has had more extensive experience than any other single operator, presents his fundamental principles:

1. Liberation of the left ventricle first, in order that it can receive and immediately deliver to the systemic circulation the increased output of the right ventricle following its subsequent decortication and thus avoid the right sided venous congestion that would otherwise occur.

2. Freeing of the right ventricle which then usually dilates more efficaciously and beats stronger. The final freeing of the right heart should be done in a systemic successive manner. Owing to the thin wall of the auricles, these should not be freed.
Whereas, previously there was considerable discussion as to the extent of the decortication, at the present time the important question is consideration of what parts of the thickened pericardium should be allowed to remain as support to the weakened heart musculature. Because of the above described danger of an acute tricuspid insufficiency and also in order to avoid the development of deficiency of the auriculo ventricular valves, thus leading to the immediate occurrence of an inflow venous congestion, the decortication should never be performed beyond the coronary sulcus.

Burwell and Blalock question Schmieden's statement that the left ventricular scar should be decorticated first. They reason that most of the signs and symptoms are attributable to back pressure from the right side of the heart and "it would seem important to decorticate the right ventricle."

Regarding the operation, White believes that the, "so-called Delorme operation of pericardial resection is the only cure for chronic pericarditis or Pick's disease. It is evident that Brauer's single operation, once called cardiolyis but better designated thoracolysis, cannot help in the least in freeing the heart from its shell of fibrous pericardium, although this operation may be successful in rare cases where there are important external adhesions gluing the heart to the chest wall itself or in patients with very large hearts, which raise the ribs in systole and not only increase their work thereby but cause much pericardial discomfort. There are four essentials for a successful operation of pericardial resection.

1. The evaluation of symptoms and signs that makes the diagnosis certain. 2. Selection of a case that is more or less crippled and yet a reasonable risk for the extensive operation that is necessary. 3. An expert anaesthetist (our own cases have had general anesthesia under ether). 4. An experienced thoracic surgeon who is bold and yet cautious, who will do enough to free the heart and yet not too much to endanger the patients life."

According to Sprague, there are four types of cases which may get relief from operation. Those with: 1. Chronic obstructing bands

We have seen that the operation of choice is the Delorme procedure of decortication of the heart. A few, however, such as Lockwood and Vaquez do not approve of the Delorme operation. Vaquez states that it is not a rational one "for isolated pericardial adhesions do not interfere with the functioning of the heart, and, even if they were destroyed, they would be reproduced eventually."

Beck, speaking on the same subject says "I believe that the Brauer operation is obsolete, inevitably, although it has served an important purpose as a stepping stone in the development of pericardial surgery. Pericardectomy, complete as is anatomically possible, is the operation of choice. It should be carried out in one stage, because the heart yields to the surgeon one major operative opportunity. If the heart is not relieved of all its mechanical impediment that operation, it may fail to recover. Before the surgeon attempts this operation on a human being he should acquire invaluable experience in its performance that the laboratory alone is able to provide."

Bigger, however, advocates a two-stage operation. He believes it is necessary to remove both epicardium and pericardium. He advises blunt and sharp dissection, with removal of small fragments of pericardium as they are freed.

Schmieden believes that the pericardium should not be peeled like an apple, but like an orange.

Regarding the vital part of the operation—that is decortication of the heart—it begins after about an hour's preparation of the field. Both blunt and sharp dissection are used to free the heart from the constricting pericardium, which may be thick and tough parietal pericardium, dense epicardium or both securely or even inseparably united. Sometimes a line of cleavage is easily found but often it is not and then great care must be used to avoid splitting the muscle itself and
one of the heart chambers, the right ventricle or right auricle in part. "However, even if the heart is penetrated the wound may be quickly repaired.....leaving a strip of pericardium attached to the heart as one dissects permits--its use as a patch of tissue to suture over the wound."10

Beck121 has demonstrated that the ability to dissect the pericardial scar from the surface of the myocardium is the procedure which may make the difference between success and failure in some cases. "This factor of epicardial sclerosis with constriction of the heart is worthy of emphasis, and improvement in technique in control of hemorrhage from sharp dissection should give better results."

Griswold90 informs us that haste has no place in this operation--that manipulations within the pericardium cause considerable cardiac irregularity and that frequent periods of rest are necessary to restore normal rhythm and maintain cardiac function within safe limits.

Regarding another problem, he tells us that the drainage of the large amount of fluid which forms about the heart after this and other cardiac operations is a serious problem. To quote, "The absorptive qualities of the mediastinal tissues are uncertain and enough fluid may collect within a few hours to produce serious compression of the heart. Leaving a drainage tract leading to the surface of the skin carries with it a real hazard of infection...The most satisfactory way out of this dilemma is to leave a generous opening (from 3 to 5 c.m.) for drainage into one of the pleura. During the post-operative period we can remove the effusion from the pleura by syringe and needle as indicated."

Any blood that is retained within the pericardium is absorbed without producing adhesion.127

If the decortication of the heart is not fully successful and the heart is bound down to the left dome of the diaphragm, then according to Schmieden and Westermann,129 "the normal systolic contraction is
impossible, because during inspiratory depression of the left dome of
the diaphragm the heart is elongated and during systole becomes lancet
shaped. In order to remedy this undesirable condition left sided phren-
ectomy is generally recommended, since this results in sufficient relief."

According to Lawrence and Morton, the main advance in the field
of surgery since 1929 "is the H shaped incision, introduced by Beck and
Griswold. By means of this operation the sternum is spared, and an
excellent exposure of both sides of the heart is obtained."

Regarding the safeguards of operative care, Churchill gives the
following suggestions: "Transfusion has not been employed and is to
be avoided because of the danger of cardiac dilation from too great a
venous return to the heart. If required, citrated blood should be
administered very slowly.

An oxygen tent is used routinely but may be discontinued in a
few days.

Drugs, other than diuretics have little place either in the pre
or post operative program.

A syringe containing adrenalin solution should be at hand during
the operation for use if cardiac standstill be encountered.

The greatest therapeutic safeguard is achieved by maintaining
adequate oxygenation both during the operation and subsequently. The
heart will tolerate many insults if adequately supplied with oxygen,
but withstands poorly any unusual strain or manipulation if attended
by anoxemia.

While Beck focuses his attention upon failure of the peripheral
circulation from a reduced filling of the heart, I consider the real
hazard of the operation to be in exposing the weakened musculat
of the heart after it has been released from the scar to too great a
venous return."
PROGNOSIS

The prognosis in this syndrome depends on whether or not surgical treatment is available. According to White,10

"The prognosis of Pick's Disease without surgical treatment is unfavorable for health and in some cases for life. It tends to be a chronic disease, lasting several to many years, and often remaining at a standstill for long intervals of time. In a few instances...the downhill course may be rapid, ending in death from the disease itself or from complications in the course of months or a year or two. Remissions may occur, especially if favored by therapy such as rest, diuretic drugs, and fluid and salt restriction, as pointed out in his own noted case by Finsen 35 years ago. Spontaneous cure probably does not exist, surgery affords the only cure, which in our experience has been effective for at least 7 years.

The occasional cases of apparent recovery for intervals of months or years after the occurrence of symptoms and signs of cardiac compression before a recurrence of persistent symptoms and signs of much the same sort are undoubtedly to be explained as having had, at first, an acute pericarditis with effusion which subsided, and later on, after healing, constriction from a chronically contracted fibrosed, pericardial sac (with or without calcification) which is in large part incidental."

The results from surgery have on the whole been excellent. When these men speak of cures—they speak of girls and boys who are now able to go out and play football and basketball and participate in other athletic contests, wherebefore a game of checkers was probably too much.35

Churchill in 192922 collected 36 cases of pericardial resection for constricting pericarditis and added one of his own. Most of Churchill's collected cases came from Germany and were reported notably by Volhard, Schmieden and Fischer. In 13 of the 37 cases the results were excellent. In 4 of the cases there was transitory improvement, but the patient died of the original disease. In 2 cases the operation was fatal, death being due directly to operative procedure. In the other 5 cases the operation was not completed.
In 1936 Churchill published the reports on 10 others. In his 10 cases he reports 6 cures, 1 death, and 3 showed marked improvement. The longest cure reported was 10 years. Blalock in 8 operations had 3 cures, 2 improvements, and 3 deaths. Stewart and Heuer, et al report similar results.

In the Massachusetts General Hospital series of 15 cases, 6 were completely cured by operation, 1 partially cured, 3 cases died of the disease itself, 3 cases died post-operative, and 2 other patients shown by signs and X-ray to have the disease, but not operated on.

The original patient operated on in 1913 by Sauerbruch, who had been confined to bed by dyspnea and showed great edema was symptom free and working 11 years later.22

Some patients show a delay in improvement after operation. This delay in improvement in certain patients after operations may be due in part to dilation of the region of the heart from which the pericardium has been resected, and in part to obstruction not having been sufficiently relieved. At operation the heart bulged through the window which was made in the pericardium and undue stretching of this muscle may have resulted. It may require time for this muscle to regain tone.

Beck12 has the same opinion, when he says, "After the compression agent has been removed by operation, the heart, like skeletal muscle, requires time to regain its normal strength. Indeed in chronic cases the compression agent is sometimes removed completely with little improvement in the circulation noticeable after operation. The venous pressure may remain elevated and diuresis may not take place for days or weeks after operation. This delay in recovery, I believe is due to the disuse atrophy suffered by the heart.

Regarding the question of new adhesions after the resection of a scar from the heart, the general opinion is that new adhesions do form, but that they do not produce symptoms12,126.

In a report of a Cabot case of Piek's disease upon which operation
had been preformed and which subsequently came to autopsy, Hallory reports;
"the pericardium was rather interesting in that the original adhesions had
together regrown. There was still no pericardial cavity: but very care-
ful examination showed that there was no sign of obstruction around the
orifices of the great vessels leading into the heart; that is the mouths
of the vena cavae and pulmonary veins were all free. I think that there
was not much evidence that circulatory failure played a significant part
in his death."

Mott believes that the prognosis depends on Broadbent's sign--
"for as the heart strength diminishes owing to degeneration, so this
sign disappears."

The prognosis is better for patients with non-tuberculous disease
according to Blalock.
The patient, a white female child, age 12, entered the University Hospital for the first time on 11-22-39 complaining of:

1. Swelling of face and eyes, then of extremities since July, 1938.
2. Intermittent spells of diarrhea since July, 1938.
4. Fever since about November 1, 1938.

The mother states that the child had always been in good health until July, 1938, at which time the above enumerated symptoms were first noticed.

The edema was cyclic. It would become progressively worse (involving face, eyelids, body and lower extremities) for about two weeks, then the child would develop diarrhea and would have 5-10 stools a day for 3-4 days. The stools at the time of the diarrhea would be thin, watery and light yellow with some mucus and of a very foul odor. No blood was noted at any time.

The child was in several hospitals without any results. The cyclic edema and diarrhea continued to date of entrance.

However, about 11-1-39 the child became more edematous than at any time previously and then developed a severe diarrhea with much cramping and nausea. At first the stools were brown but after about 1 week with the bowel movements as frequent as every three hours, they became tarry black in color (described by mother as resembling meconium). At this time the child began to complain also of a sore mouth and tongue and the mother states the child had sores inside of her lips and her tongue and the throat was bright red and very sore.
Throat--The mucosa of the mouth and throat shows the presence of numerous partly healed ulceration. The tongue is smooth, slightly reddened and there were a few large papillae scattered over the surface.

Heart--Neg. B.P. 112/78.

Lungs--Complimental spaces cannot be definitely demonstrated.

Abdomen--Considerable enlarged and gives the appearance of being ascitic. No definite fluid level can be demonstrated. There is a vague mass in the RUJ which is rather firm and cannot be definitely outlined because of the tenderness of the abdominal wall--possibly liver. There is tenderness on deep palpation over this mass.

Laboratory:

Blood--11-23-39, Hb 73%, RBC, 4,070,000; WBC 9,250; Seg, 24; st.68; L, 7, M. 1.

Urine--negative except for slight trace of albumin.

Blood NPN--29.7 mg.%

Total serum protein, 5.8 mg.%

Total serum albumen 3.0 mg.%

Total serum globulin 2.8 mg.%

Platelet count, sedimentation rate, blood sugar, blood chlorides, blood cholesterol and fragility of red cells within normal limits.

Kidney function tests within normal limits.

Sed. Rate 1 cc settled time Linsemmeier 6 mm--30' 12 mm-48'

18 mm--1 hr-10' 24 mm--2 hr. 10'

12-2-39 Brom Sulphalein Test

5 min-----70% dye retention

30 min-----15% dye retention

11-27-39 Flat plate of abdomen. Diaphragm appears elevated. Studies
show fluid in peritoneal cavity. EKG showed some myocardial damage.

Course in Hospital:

Daily fluid intake was from 1000 to 1500 cc. Daily output averaged around 200 cc.

Upon admission temperature was 100'. On 11-24-39 it was 103' after 11-25-39 patient began to have a gradual elevation of temperature from a low level at noon to a high level at 8PM when it was 103'. Respiration around 20 a minute. After 11-24-39 reached peak of 50 per minute which is highest at midnight. Pulse averaged about 110. Patient received Thiamin chloride intravenously, Percormorph oil, Corelexin, and high protein, low salt diet.

On 12-9-39 patient's temperature was elevated to 104.2' R in the afternoon. Examination of the chest revealed moist rales heard throughout both lungs. Respirations were rapid (50 per min) and labored. Patient irrational at times. Aspirin grs. V. every three hours ordered. Respirations ceased suddenly at 10:47 P.M.

From the record of the patient at the Methodist Hospital where she was from 7-1-39 to 7-9-39 we have the following additional information:

The scarlet fever had an ordinary course without complications or noticeable sequelae.

In the history we find the statement that "patient has been under medical care almost constantly. Treated for heart disease. Whole breast seemed to hurt with her heart."

Lungs: Faint subcrepitant rale in both bases, no dullness. Excursion 2 cm. Heart sounds clear, occasional extra systole. BP 108/38.

Of four urine samples run only one showed a trace of albumin.

Kline and Kahn negative.

N.P.N. 33.3 mg%
Total Serum protein 5.3 mg%
Total Serum albumin 2.7 mg%
Total serum globulin 2.6 mg%

X-ray

Posterior-anterior radiographic study of the chest shows the pleural sacs and pulmonic fields to appear essentially clear. The broncho-vascular markings are slightly thickened about each hilar region but there is no definite evidence of a pneumonic process. No gross amount of fluid is present in either pleural sac. The heart appears to be normal in size, contour, and pulsation.

BMR - (minus) 26%

Admission note gives information that child has been on salt free diet for 6 months. Has had rapid tooth decay since PI.

EKG Flat and depressed T Wave in Leads II III IV.

Clinical conclusion myocarditis. While child was in Hosp. ran a remittent type of fever. Highest about 2 PM and lowest about 4 AM. The high was about 100'.

Autopsy Report:

External examination--The body is that of a well developed and rather poorly nourished child of about the stated age. The abdomen is distended. There are puncture marks in ante cubital fossa; and some edema in the lower extremities.

Primary incision--usual Y shaped.

Panniculus--practically none.

Peritoneal cavity--there is considerable free gas. The peritoneal cavity is divided by firm adhesions into two cavities at a level of about 3 cm. below umbilicus. Small bowel is matted together to form
this partition. The upper cavity contains much greenish, yellow pus--
cultures were taken--, the lower cavity shows fecal contamination. This
lower cavity passes upward along the right lateral body wall to a cavity
just inside the rib margin where there is a communication with the bowel.
The liver marks part of the upper margin of this last named cavity.
Pleural cavity:
Right--Some fibrinous adhesions. Practically no fluid.
Left--About 75 cc clear fluid.
Pericardium:
  Cavity obliterated completely by firm fibrous adhesions. Peri-
  cardium thickened. Occasional spots of calcification between the two
  layers of pericardium.
Heart:
  Weight 140 grams.
  Heart muscle soft.
  Tricuspid valve--no change.
  Pulmonary valve--no change.
  Mitral valve--no change.
  Aortic valve--no change.
Lungs:
  Right--weight 310 grams. Large amount of frothy fluid in bronchi.
  No changes in main branches of the pulmonary artery. Few small masses
  of ante-mortem thrombi the vessels. Cut section edematous and shows
  extreme consolidation inferiorly and posteriorly.
  Left--weight 280 grams. Left lung and bronchi and blood vessels
  same as on right side. Cut surface also similar except for some areas
  of necrosis in the base.
GI tract and esophagus--normal.

Stomach--Dilated, mucosa is thin and shows little other changes.


Small bowel--Considerable hypertrophy.

Large bowel--Two small perforation about 6 cm above the iliocecal valve, with considerable inflammatory reaction of mucosa surrounding this area.

Liver--Weight 1310 grams. Surface is quite irregular and granular. The inferior surface of the right lobe is discolored. Gall bladder wall is considerably thickened and its mucosa is quite normal in appearance. Cut surface of liver shows considerable accentuation of the markings. The whole tissue is rather pale.

Spleen--Weight 140 grams. Cut surface pale, tissue soft and markings are quite indistinct.

Pancreas--Seems to be somewhat enlarged but section shows a normal appearance. Lymph nodes about pancreas are greatly enlarged.

Kidneys--Left--Weight 100 grams. Smooth in contour. Soft; markings distinct. Right--Weight 100 grams. Entirely similar to left.

Adrenals--Both appear normal.

Pelvic organs--Tubes and ovaries are bound down by numerous firm adhesions so that individual structures cannot be identified in the peritoneal surface.

Urinary bladder quite smooth and shows only a few petechia on its mucosa. Uterus is infantile in type. Ovaries are small and contain only an occasional small follicle.
Summary:

1. Chronic peritonitis (probably pneumonic).
2. Perforation of colon.
3. Chronic adhesive pericarditis.
4. Early cirrhosis of liver.
5. Pulmonary edema.

(Cultures taken didn't grow. Stained slides show pneumonia.)

Microscopic examination:

Heart--Hyaline thickening of epicardium with small patches of calcification. Pericardium thickened.

Muscle layers are split by edematous fluid. There are patches where one finds individual hypertrophy of muscle fibers. Other areas show degeneration of muscle fibers. Cloudy swelling of the muscle fibers is also seen. There is some increase of connective tissue.

Lungs--Alveoli are congested. Alveolar walls are infiltrated with edematous connective tissue in which there are scattered areas of polymorphs and monocytes.

Liver--The liver cords are pale staining. The cells show vacoules and cloudy swelling. There is some increase of interlobular and portal connective tissue. The sinusoids are dilated.

Spleen--Normal in appearance.

Pancreas--Normal in appearance.

Kidneys--Severe cloudy swelling of the tubules.

Adrenals--Normal in appearance.
Lymph node--Isolated patches of polymorphs and plasma cells and dilated capillaries.

**Summary of Microscopic Examination:**

1. Chronic constrictive pericarditis of unknown origin.
2. Congestion and edema of the lungs.
3. Cloudy swelling and beginning cirrhosis of the liver.

**Final diagnosis:**

Picks' Disease with polyserositis.

This case was labelled at autopsy, for want of a better diagnosis as Picks' Disease with Polyserositis. The redeeming feature is that it was recognized at the time the label was applied that it was not a true case of Picks Disease. However, diseases much more remote from what White, Churchill, Blalock and Back would recognize as Picks' syndrome have been given that diagnosis with much less provocation.

The glaring omission is want of a statement that there was dilation of the neck veins. We may assume that there was no dilation because many good men examined the child and did not note such. Would we not assume that if the edema of the face was the result of "constriction of the superior venae cavae as it passes through the pericardium" that we would also have venous engorgement of the neck and face? The so called superior mediastinal syndrome of Kreisle and Hardinick.130

Another factor present in this case is the intermittent spells of diarrhea and the cyclic nature of the edema. As far as I have observed diarrhea is usually not associated with the Picks' syndrome. In only one case--that presented by Cabot in 1898 did I find diarrhea mentioned
in the symptoms. Dr. Tremaine\textsuperscript{13}, however, presented two cases of what he calls Pick's disease of the subacute type with a history of acute diarrhea, nausea and vomiting. One of the cases is interesting to compare with the one presented here, because of the demarcated red areas found in \textsuperscript{15} on the child's body and the ulcerations found in the child's mouth upon examination.

The case in question was a girl, age 19, who developed non-pruritic, reddish purple, maculo-papular eruptions on the face, neck and behind the ears. Also, numerous vesicles on hard palate and buccal mucosa. The buccal lesions became ulcerated and covered with a grayish membrane. Within two weeks the rash had disappeared followed be scaling, but leaving a dusky complexion but no scarring or atrophy. At autopsy the outstanding features were polyserositis, nephritis, and the absence of rheumatic myocarditis or endocarditis. The polyserositis was characterized by the following findings. An obliterate fibrous pericarditis with adherence of the pericardium to all of the surrounding structures; a fibrous pleuritis so extensive that the lungs were adherent to the parietal pleura over the greater part of their surface; fibrous adhesions which were most numerous in the upper abdomen where they completely embedded the liver spleen, and transverse colon and also involved various segments of the small and large intestine throughout the lower half of the abdominal cavity; inflammatory changes of the synovial membrane of the right knee joint. Evidence of continuance of infection was shown by mild infiltrations with lymphocytes and plasma cells and occasionally polymorphs throughout the fibrous adhesions and by the presence of a few small fibrin masses and fibroblasts that were still laying down connective tissue. Autopsy also showed kidney damage and
The heart examined by radiographic methods showed no abnormalities; pulsations were noted and not found to be unusual—yet at autopsy, we find a pericardium which certainly might have interfered in the main with normal pulsations. This is one part in favor of Pick's syndrome. Yet we find hypertrophy of the individual muscle fibers along with atrophy at autopsy. Can this be explained as suggested by Dr. Tollman that the degeneration of some fibers have left room for the others to hypertrophy in? Can we explain it on the basis of areas of the heart not being completely constricted—the heart at autopsy showed the scar tissue not to be of universal thickness—and these areas taking on the workload of the rest of the heart.

The blood pressure is not characteristic of Pick's disease. It is recorded as 112/78 at the University Hospital and at the Methodist as 112/38.

With the history of fever since only 11-1-39 we could have worked out a nice theoretical history of how on about this date, because of the lowered resistance of the peritoneum by the decreased blood flow and venous stasis, pyogenic organisms got a foot hold and chronic peritonitis resulted. From a time body defense and invading organism which was probably a pneumonocci were of almost equal strength, but after a time the pneumonocci gained control with the result that an abdominal picture resulted as we saw at autopsy. A similar idea is expressed by Reed in explaining how Fungi complicated a case of Pick's disease.

Locus minoris resistentiae rolls nicely off one's tongue, but it is somewhat Don Quixotish.

It does not explain to satisfaction the case in question when we consider that there was a remittent type of fever in July of 1939, when
the patient was at the Methodist Hospital. How long previous to this
did the child have a fever? Could we account for the high afternoon
temperature on the basis of an atypical form of tuberculosis?

The serum proteins of the blood presented a not unusual picture.
We do at times find a low serum protein in Pick's disease.

Flick and Gibbon\textsuperscript{132} believe that "the importance of determining the
serum proteins in patients with Pick's disease is obvious. If the
protein content is low a second factor is brought in to play in the
production of the edema and ascites which may as well be as potent as
the increased venous pressure."

Strong\textsuperscript{133} thinks that the reversal of the normal serum albumin,
globulin ratio is probably only evidence of albumin loss, due to re­
peated aspiration. He also thinks that it is due to damage to the
liver cells.

In the present case we may assume that the serum protein is on
the basis of low protein intake and also the basis of the diarrhea.

An avitaminosis was present clinically as seen by the physical ex­
amination of the mouth.

The liver as seen grossly and microscopically with the beginning
cirrhosis is a point in favor of calling the condition Pick's disease.

In the final analysis we are confronted with the problem of whether
or not the condition of the heart as seen at autopsy was sufficient to
account for the patients symptoms and whether surgery would have eli­
minated the symptoms. We are somewhat handicapped by the fact that
Pick's disease was not considered at this time—and as a result our in­
formation regarding encroachment upon the Vena Cavae is meager. The
normal pulse pressure, and the radio graphic studies would tend to
indicate that the heart was not under much of a constrictive handicap.

It is of interest to list the points in favor and against a diagnosis of Picky disease:

<table>
<thead>
<tr>
<th>For</th>
<th>Against</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Thickened pericardium with adhesions and calcification.</td>
<td>1. Lack of dilation of the neck veins.</td>
</tr>
<tr>
<td>2. Beginning cirrhosis of the liver.</td>
<td>2. The low B.M.R.</td>
</tr>
<tr>
<td>3. Lack of response to medication.</td>
<td>3. Normal pulse pressure and radiographic findings.</td>
</tr>
</tbody>
</table>

**Questionable Facts**

1. Ascites with preponderant edema of a cyclic nature with diarrhea.
2. Beginning reversal of the serum albumin, globulin ratio.
3. Polyserositis.

If Picky disease is to mean a condition in which the symptoms are primarily a result of pericardial constriction, I don't think the label fits here. Operation in this case might have relieved the compression temporarily, if any were present, but I doubt if her life would have been prolonged.
CONCLUSIONS

1. Picks' Disease or Syndrome is as good a term as any to describe the symptom complex which on a physiological bases is the result of an inflow stasis. It is a broad and vague term and is often used incorrectly - but until more is known regarding the patho-anatomical ramifications - we have none better.

2. The etiology of this condition is not known - a rheumatic basis is quite rare and tuberculosis a quite common finding.

3. An effort should be made to differentiate this condition from those diseases which produce a similiar picture because of the possibility of a surgical cure.

4. There are many helpful diagnostic aids - the most valuable of which are measurement of venous pressure and radiographic studies of the heart.

5. Surgery should be selective and only attempted by those with previous experience.
BIBLIOGRAPHY


43. Tice, Frederick, "Diseases of the Pericardium." Practice of Medicine, pp. 225-235.


73. Frosch, Herman, "Picks Syndrome." New York State Journal of Medicine 38 (Pt. "2"):1186-1188, Sept. '38.


78. Stewart, Harold, and Hauer, George, "Chronic Constrictive Pericarditis—Dynamics of the Circulation and Results of Surgical Treatment." Archives of Internal Medicine, 63:504-530, March '37.


92. Schwab, Edward H., and Herrmann, George, "Alterations of the Electrocardiogram in Diseases of the Pericardium." Archives of Internal Medicine, 55:917-941, June '35.


108. Vaquez, H., Diseases of the Heart, Pp. 239, Translated by George Lidlaw, W. B. Saunders Co., Phil. 1924.


