THE PATHOLOGY OF SYphilis*

With Special Reference to the Development of Luetic Aortitis

Harrison S. Martland

Newark, N. J.

The pathology of syphilis of the circulatory and nervous systems will be discussed from the viewpoint of a general pathologist, outlining the anatomical pathways of infection in acquired syphilis.

Most of the facts herein stated are generally accepted by recent authorities and are based upon clinical, pathological and experimental evidence.**

I shall draw from these some deductions and conclusions which may point the way to further research into, and increased understanding of this disease.

INOCULATION AND PRIMARY INCUBATION PERIOD

The treponema pallidum is usually deposited on an abrasion on the genitals. An abrasion is, however, not necessary and it may often be microscopic.

The spirochetes on account of their motility and also probably due to the capillary attraction of the open lymph-spaces are carried deep into the initial site of inoculation and there is a rapid reproduction of the organisms in the lymph-spaces and perivascular lymphatics.

Many pass rapidly into the capillaries and reach the important viscera within two to three days after inoculation. Hence, so-called abortive excisions of the chancre and abortive injections of arsphenamines, etc., will not prevent the development of syphilis.

*Delivered before The New York Academy of Medicine, March 3, 1932.

**In preparing this sketch, I have quoted extensively from the writings of Stokes, with whose views I am in entire accord.
Syphilis is, therefore, a systemic general infection days and even weeks before the appearance of the chancre.¹

The greater number of the rapidly reproducing spirochetes, however, enter the surrounding lymphatics and drainage on the slower lymphatic stream to the regional lymph-nodes begins.

**Primary Stage—Chancre**

As the rapidly growing spirochetes increase in number, the endothelial cells of the capillaries swell and an obliterating endarteritis occurs. Many new capillaries are formed. Perivascular collections of lymphocytes and plasma cells appear at an enormous rate and a papule with induration results. The typical Hunterian chancre, a local defense reaction at the site of inoculation, is formed.

Spirochetes rapidly reproduce and the initial sore teems with organisms. The direct feeding of the blood stream continues and increases, producing a bacteremia. Multiple, metastatic foci develop throughout the body. Their number, location and activity are proportional to the virulence of the organisms and the host.

The feeding of the slow lymphatic stream increases. Spirochetes reach the regional lymph-glands. Many are killed by the phagocytic activity of the reticuloendothelium of these nodes but some always survive and pass these defensive reservoirs and eventually reach the blood through the lymphatic duct, etc., or remain locked up in the lymph-nodes, to reinfect the lymph-stream or blood years after.

Clinically, during this stage, the chancre develops with wide variations in size and appearance. The local lymphadenitis develops with satellite bubo. Systemic symptoms, such as headache, pains in bones, etc., are frequent and even occur before the chancre. The blood begins to show a positive Wassermann reaction.

**Early Secondary Stage**

The chancre now heals with the production usually of a tell-tale scar. Spirochetes become fragmented and scant.
Lymphocytes and plasma cells disappear and fibrosis takes place.

The remaining organisms are usually completely destroyed or they may become locked up in the fibrous scar and form the nidus of a chancre redux.

Many of the early metastatic foci are now healing, in a manner exactly similar to the healing of the chancre. Local immunological reactions undoubtedly play an important rôle.

The marked bacteremia from the chancre and early secondary foci causes countless secondary infections of the skin, bones and important viscera. In these areas of infection a rapid reproduction of spirochetes takes place followed by a characteristic cellular reaction, similar in every respect to that of the original chancre, with consequent formation of innumerable secondary lesions.

Clinically, the secondary rash appears. Macular, if the predominant feature is the formation of new capillaries, and papular if the perivascular collections of lymphocytes and plasma cells are marked.

A generalized lymphadenitis occurs, due chiefly to lymphatic drainage from the innumerable lesions of the skin and mucous membranes. Enlargement of the spleen is common. Bone and nervous lesions begin. Special structures, as the eye, ear, liver and kidneys may be affected with serious results. A positive Wassermann reaction develops with the refractory state.

**Late Secondary Stage**

Systemic defense, with the production of agglutinins, lysins, etc., now comes into play. Systemic resistance reaches its highest point converting the disease into latent syphilis.

Secondary foci in all stages of activity and decline occur all over the body. Healing occurs by fibrosis. Most of the spirochetes are destroyed. Sterilization, however, is never
complete and provides bases for relapses, which are so characteristic of all stages of syphilis.

Spirochetes disappear from the blood, or occur in occasional showers.

In no stage of syphilis do the spirochetes multiply in the circulating blood to any appreciable extent.

Parenchymatous tissues, such as the lungs, heart muscle and liver, rid themselves of spirochetes by drainage into the regional lymphatics and lymph-nodes.

The spirochetes enter the perivascular lymphatics, the lymph-node reservoirs and inaccessible parts of the bones and nervous system, where they cannot be reached by the circulation. Here they rest, or reproduce in the quiet, mild, anaerobic lymph. Syphilis now becomes a disease of the lymphatics and ceases to be a blood disease, except for occasional showers of spirochetes, usually scant in number, which may be thrown into the blood stream due to the re-awakening of old microscopic foci.

Clinically, in this stage, the secondaries disappear spontaneously. Systemic manifestations subside. The only clinical evidence remaining being a palpable spleen and liver and asymptomatic neurosyphilis.

Due to defects in resistance or revival of the partly extinguished foci, any lesion in primary or secondary stage may reappear, especially those on the mucous membranes.

**INFECTION AND DRAINAGE OF CERTAIN VISCERA**

The mode of infection by the blood stream and the manner of the riddance of spirochetes from some of the viscera in the early and late secondary stage is of great importance, and must be considered in more detail.

**THE LUNGS**

In the stage of bacteremia when the chancre is feeding the blood stream with innumerable spirochetes, the lungs are the first important organs to receive the dosage of or-
ganisms. In fact, on account of their very great capillary area, they receive a greater dosage than perhaps any other part of the body, with the exception of the skin.

The lungs are the most important and extensive capillary filtering bed of the venous circulation for large sized particles.

Let us take for example, a case of septic endometritis, complicated by a thrombophlebitis of an ovarian vein. Constant showers of hemolytic streptococci, mixed with blood platelets, fibrin, polymorphonuclears and agglutinated cocci enter the inferior vena cava, pass rapidly through the right side of the heart and enter the lungs. Most of the streptococci are filtered out or retained in the lungs and multiple, symmetrical, bilateral, hematogenous, embolic abscesses occur. Infection of the pleura by continuity, or by rupture of a surface abscess, with the production of an empyema often ensues. The lungs in such cases show a remarkable ability to rid the blood stream of the infection. Sometimes the patient may even recover. If she dies, there may be little evidence of systemic emboli.

In the bacteremia in the second stage of syphilis, however, the spirochetes are unaccompanied by large size particles, such as agglutinated bacteria, etc., and it is reasonable to suppose that most of the spirochetes easily pass through the lung capillaries and enter the systemic circulation.

Many, however, are undoubtedly slowed in the lung capillaries, cling to the side of the vessel walls and, because the treponema pallidum are always attempting to leave the circulation for extravascular tissues, in which they are less liable to injury, penetrate into the interstitial portions of the lung.

Those that do penetrate the lung parenchyma, in their attempt to get far away from the blood stream, will not under ordinary circumstances remain there for any length of time. First, they do not like the proximity to oxygen; second, many will be phagocyted by the alveolar epithe-
lium, which is mesothelial in origin and a part of the reticuloendothelium; finally, and what is more important, the spirochetes will be taken up rapidly by the lymphatics and drain into the hilus nodes, due to the great drainage power of the lungs in ridding themselves of foreign and undesirable particles.

A similar process is seen in the handling by the lungs of the tubercle bacillus, in the storage of coal pigment and silica in the hilus nodes and in the remarkable lymphatic drainage seen in malignancy of the lung.

The lungs eventually rid themselves of the spirochetes by way of their lymphatics, which drain to the peribronchial and regional nodes of the mediastinum. These become collecting stations, or reservoirs, for the storage of spirochetes. Here they are exposed to the phagocytic action of the reticuloendothelial cells of the lymph-sinuses.

I am of the opinion that the above conception explains the rarity of pulmonary syphilis. It is a notable fact that in acquired syphilis, gummata of the lungs are unusual. The various forms of pulmonary syphilis described by clinicians and roentgenologists and seldom substantiated at autopsy, have usually been of the hilus type. A diffuse, fibrotic infiltration forms a doubtful and nebulous pathological entity.

Furthermore, it is of interest to note, that the only common form of lung syphilis is that seen in stillborn infants, or in those who live only a few days, in which the well known "pneumonia alba" is encountered. Microscopic examination of these lungs show chiefly an arrested development rather than an active inflammation, with considerable cellular fibrous tissue about the blood vessels.

In heredosyphilis the child is infected through the placenta, and an incalculable number of spirochetes circulate throughout most of the organs, the number having no parallel in acquired syphilis. The rarity of heart disease in congenital syphilis might be explained, therefore, by the fact that the lungs of the fetus have escaped the large
dosage of spirochetes, because the fetal circulation does not pass through them to any great extent.

In instances in which the lungs receive an unusual dose, a fatal pneumonia alba occurs, principally because the lungs are not aerated and offer a more favorable soil for the reproduction of spirochetes than do the adult, oxygen containing, lungs.

**THE HEART**

In the septicemia stage, spirochetes undoubtedly reach the heart muscle in considerable numbers by way of the coronary circulation.

Does syphilis of the heart muscle begin in this stage?

Warthin's observations led him to conclude that in the second stage of syphilis there is a very active specific infiltration of the heart muscle.

Clinically, it is well known that various cardiac disturbances take place during the early stages of syphilis, such as, tachycardia, bradycardia, arrhythmias and syncope. It is doubtful, however, whether these are due to a true specific myocarditis.

It is obvious that this question is very important. If there is a specific myocarditis in early syphilis, there is a foundation laid for the various forms of myocardial syphilis said by Warthin to occur in the later stages of the disease. (Active syphilitic myocarditis, chronic syphilitic myocarditis and fibroid myocarditis with acute exacerbation).

I am of the opinion that the heart muscle is not favorable soil for the growth of treponema pallidum. Because of the firm, ceaseless contractions, it is too active a place. The oxygen content is too high. I believe the strong muscle contractions favor lymphatic drainage towards the aortic arch and help in ridding the myocardium of the organisms.

Those remaining and surviving may rest for many years without doing appreciable damage. The finding, therefore,
of a spirochete in the heart muscle in late syphilis, perhaps surrounded by a few lymphocytes, may mean nothing more than an insignificant historical landmark. In a similar manner spirochetes may often rest in the skin of latent syphilitics, leaving no gross, and little microscopic evidence of their presence. Yet syphilis may be transmitted to rabbits by inoculation.

It is of interest to note that very few have ever been able to find spirochetes in the myocardium in acquired syphilis, although most careful and painstaking technic has been used. Furthermore, in using the Warthin-Starry technic, it has been repeatedly shown that artifacts indistinguishable from spirochetes may be obtained by staining cover slips alone, without any tissue.

Saphir\(^{6}\) in a review of the recorded cases of syphilitic myocarditis in literature states: “A critical consideration reveals that morphologically the diagnosis of syphilitic myocarditis cannot be made in any of the reviewed cases without the presence of gummas.” Furthermore, perivascular infiltrations of lymphocytes in the myocardium; lymphocytes, plasma cells and endothelial cells in the interstitial tissue; and polynuclears may be found in the myocardium in cases of coronary sclerosis especially in cases in which there has been infarction and in cases of rheumatism and other infections. They are not pathognomonic of syphilis.

**THE NERVOUS SYSTEM**

In the stage of spirochetemia, innumerable spirochetes reach the brain by way of the carotid circulation.

Many of these eventually reach the pia-arachnoid, chiefly over the base of the brain. Reproduction is followed by the characteristic, cellular reaction.

Even as early as the stage of the chancre, an increase in the cell count of the spinal fluid may occur, denoting the existence already of an exudative process in the leptomeninges.

In the secondary stage, spirochetes have been found in the spinal fluid, both by inoculation and the microscope.
THE PATHOLOGY OF SYPHILIS

While infection of the nervous system is common and occurs early in the disease, the actual development of syphilitic disease is much less so, and only around 20 per cent of all syphilitics show during their disease meningo-vascular symptoms.

That all syphilitics do not develop neurosyphilis, tabes or paresis is due to many factors, such as dosage, resistance of host, immunity, and possibly differences in strains of organisms.

The common location for early neurosyphilis is over the base of the brain, where when the lesion is visible, a gelatinous exudate is seen over the interpeduncular spaces, or a milkiness so slight that it may easily be overlooked. This location accounts for the early involvement of the third, fourth and sixth cranial nerves with symptoms of diplopia, strabismus, etc.

The manner in which the spirochetes gain access to the subarachnoid spaces over the base of the brain is of great interest.

In tuberculous meningitis the tubercles are located, chiefly, in the Sylvian fissures along the course of the middle cerebral arteries. For years they were interpreted as hematogenous in distribution, springing from the vessels of the pia-arachnoid. While this appeared plausible, it has for many reasons, never satisfied critical pathologists.

Rich4 then demonstrated, experimentally, that the meninges could not be infected by direct injections of tubercle bacilli into the carotid arteries, but typical tuberculous meningitis could be produced by subarachnoid injections. He regarded a primary infection of the choroid plexus, a hematogenous one of course, as the essential factor in the production of tuberculous meningitis, the bacilli entering the interventricular fluid and finally reaching the subarachnoid spaces over the base of the brain.

Lewkowicz5 called attention to the same mode of infection in meningococcus meningitis.
I am of the opinion, that the same anatomical route occurs in syphilis, the spirochetes entering the lateral ventricles by way of the choroid plexuses. Because of the large size of the ventricles, their smooth walls and the downward flow of spinal fluid, the spirochetes are rapidly washed out of the ventricular system into the basal cisterna where they finally lodge in the quiet and restful interstices of the subarachnoid spaces over the base of the brain.

Such a conception best explains the location of early neurosyphilis and the logical development from it of meningoencephalitis and general paresis. In paresis, the granular ependyma of the lateral and fourth ventricles is pathognomonic, a heaping up of neuroglia proliferation probably due to spirochetal rests which have been caught in the ependyma.

Such a conception warrants careful histological study of the choroid plexuses in this disease, which seem to have been overlooked.

**LATENT STAGE**

In the latent stage of syphilis, the organisms are held in check by the local and systemic defense, the innumerable, minute, imperfectly healing foci producing immunity.

Local foci may flare up at any time, discharging a shower of spirochetes into the circulation, or into a lymphatic drainage area. Reinfection takes place, followed usually, by reproduction, typical lymphocytic cellular reactions, with consequent fibrosis and suppression of the infection. In many of these foci all the spirochetes are not killed and relapses occur.

This flareup of partly extinguished foci with consequent imperfect healing and vascular changes, results in damage to the surrounding tissues and parenchyma and the most dangerous stage of syphilis, the degenerative phase begins.

The clinical picture of this latent stage is usually characterized by prolonged absence of symptoms. Relapses are
characterized by fewer, more localized and destructive lesions due to the pronounced vascular changes and possibly to allergy. Spirochetes are scant in number and the patient is clinically not infectious.

**Late Syphilis—Gummatous Stage**

In late syphilis, due to the allergy or hypersensitiveness, and especially to the chronic vascular changes, gummas are prone to develop.

Gummata are gummy, tumor-like masses varying in size from those just visible to larger masses several inches in diameter. They are not to be confused with the miliary gumma. They may occur in any organ or tissue.

They contain very few spirochetes and the lesion is not clinically infective. They are circumscribed and composed of granulation tissue with a center of caseation due to ischemia produced by the obliterating endarteritis. There are collections of lymphocytes and plasma cells around the center of caseation, together with a few giant cells. On the edge, proliferating fibroblasts and a definite fibrous capsule may be noted. The result is extensive sloughing with destruction of parenchyma and enormous scarring.

On account of their size, gummata usually produce symptoms and are easily diagnosed. They are consequently becoming less frequent.

Gummas are more likely to be encountered in the brain, where on account of their size, they must be differentiated from other cerebral tumors; in the calvarium, where they cause extensive destruction of the skull, fortunately infrequently seen, nowadays; in the larynx, where they produce hoarseness and sometimes cause death from asphyxiation; in the mediastinum, where they represent the only common form of thoracic syphilis; in the lung, where they are infrequent; in the liver, where they lead to extensive destruction with the production of the well known hepar lobatum; in the heart muscle, where they are rare, being only seen in large autopsy services and usually occurring on the in-
terventricular septum; in the testes, where they have often been erroneously removed for malignancy; in the rectum, where they produce strictures; in the bones, commonly in the tibiae, where they must be diagnosed from other bone tumors; and in the skin, usually over the legs, where they may produce extensive ulceration and scarring.

**Late Syphilis—Degenerative Stage**

Syphilis is frightful, not because of destructive lesions of the skin and bones, but because of the late, insidious complications.

From 10 to 15 to 20 years after the infection, when the patient has forgotten all about his disease, after years of apparent health, and at the age when he is carrying his heaviest responsibilities, he is suddenly confronted with the fact that he has been gradually developing a serious, irreparable, and eventually, fatal disease of his nervous and cardiovascular systems.

This degenerative stage is the result of the imperfect healing of the innumerable, metastatic, miliary gummatas which occur throughout the disease, in any organ or tissue of the body.

The small, microscopic perivascular collection of lymphocytes and plasma cells forming the miliary gumma is the characteristic histological unit of syphilis, as is the tubercle in tuberculosis and the submiliary tubercle in rheumatic fever.

Due to imperfect healing and incomplete sterilization there is a constant flaring up of these foci throughout the disease. New adjacent and far distant foci are formed by reinfection. Reproduction, a cellular defense reaction, fibrosis and imperfect healing take place. In time this vicious cycle causes irreparable damage to the parenchyma and surrounding body tissue. Serious disease, especially of the nervous and cardiovascular systems, results.

In the nervous system, the early neurosyphilis, if not cured, may become markedly meningoencephalitic in type
and paresis develops. In this disease, the spirochetes become very numerous, unlike that in any other late lesion in syphilis. Or tabes may develop, usually due to this same gummatous infiltration of the posterior ganglia, the changes in the posterior columns being entirely those of secondary degeneration. Hence, spirochetes are very scant in the cord.

**SYPHILIS OF AORTA AND HEART**

In syphilis of the aorta and heart the main lesion is usually in the first portion of the aortic arch.

During the stage of spirochetemia, we have already noted the countless metastatic foci established in the skin of the entire body (as manifested by the rash), in the mucous patches in the mouth and pharynx, and over the genitals. About each focus, reproduction takes place, characteristic cellular reactions and then subsidence and healing. On healing the remaining spirochetes drain into the regional lymph-nodes. From the face, mouth, throat, chest and upper extremities the final drainage must be into the mediastinal nodes.

Furthermore, this is augmented by the drainage into these same nodes of the spirochetes coming from the lungs.

The hilus and mediastinal nodes then become, in fact, the largest and most important group of nodes draining spirochetes from the parenchymatous organs and tissues and the largest collecting stations or reservoirs for the storage and destruction of spirochetes.

The lymphatic network leading to these glands and away from them is enormous. The moving of the parts during respiration, greatly increases the possibility of retrograde lymphatic permeation and embolism. Although some pathologists ridicule retrograde lymphatic drainage, it is a well established mode of metastasis in cancer.

The most important retrograde extension from these nodes would be to the first portion of the aorta.

The main and most important lesion in nearly all cases
of acquired syphilis of the aorta is a supravalvular sclerosis. Most of the other changes and phases of cardiac syphilis depend upon this lesion.

The earliest lesions are microscopic and occur around the vasa vasorum in the adventitia of the root of the aorta where there is seen a collection of lymphocytes and plasma cells, lying, probably, in the perivascular lymph-spaces. Stained sections may show spirochetes in these areas, but usually they are found with difficulty. Small, miliary gummata are formed in the adventitia.

There follows a secondary invasion of the media with consequent breaking up of elastica and weakening of the vessel wall. Obliterating endarteritis of the vasa vasorum is common. The lesion develops to a stage at which it can be recognized with the naked eye. The earliest patch is usually triangular and situated just above the commissures distal to the attachment of the aortic cusps. The base of the triangle is usually pointed upward. There is a gray or slightly yellowish elevation with steep edges, rubbery and smooth on top, but marked by shallow, parallel or stellate furrows.

The process spreads in a horizontal manner around the root of the aorta (girdle of Venus). Distally it often extends to the mouths of the great vessels springing from the aortic arch. The orifices of these arteries may often be narrowed to a marked degree. The sclerosis may diffusely involve the whole aortic arch and extend into the thoracic aorta.

Histological study shows that the gummatus process starting and most pronounced in the adventitia, has infiltrated the media, breaking up and pushing aside elastic fibers, and has gained access to the subintimal tissue of the aorta where there is less resistance to further infiltration.

I speak of the whole process as a sclerosis because the lesion found at autopsy is in the nature of a deforming
scarring defect, due to the insidious gummatous infiltration, with imperfect healing and fibrosis.

Acquired cardiac syphilis is, therefore, essentially a supravalvular sclerosis which may manifest itself in one or more of the following ways:

The first, most frequent and most dangerous is the production of an aortic regurgitation. In my series, aortic regurgitation occurred in about 60 per cent of syphilitic aortitis which result in death (combined hospital and medical examiners' cases), and in 36 per cent of cases of sudden death due to syphilis of heart and aorta.

The regurgitation was always a secondary extension from a syphilitic aortitis and usually occurred as the result of a descending process. Extension of the syphilitic gummatous process through the media into the subintimal spaces producing the triangular patches, continues in the direction of the attachment of the aortic cusps, usually following the lines of least resistance, which is alongside of and between the fan-shaped subintimal fibers, the remnants of those fibers forming originally the aortic cusps.

The result is a pushing apart of the cusps at their attachments by the gummatous and sclerosing process. Sometimes the attachments of the cusps may be separated by at least 1 cm. Often, just a furrow exists between the attachments of the adjacent cusps, which are thickened and infiltrated. This widening of the commissures is the earliest sign of aortic regurgitation, and is practically the main factor in its production.

The aortic valve may also be attacked by the gummatous process ascending through the sinus of Valsalva. In this process only the aortic layers of the cusps will be involved.

The syphilitic lesion does not as a rule reach the free edges of the valves. The characteristic thickening and rolling of the free edges of the cusps in syphilis is a functional adaptation to the primary dilatation of the aortic ring.
and the thickening is a functional marginal sclerosis made worse by the mechanical effect of the regurgitant blood stream. The endocardial thickenings on the interventricular septum below the cusps is also mechanical due to regurgitant stream.

The aortic regurgitation is often combined with narrowing or atresia of the coronaries, and it is sometimes superimposed upon an old aortic aneurysm.

The heart nearly always shows marked hypertrophy and often dilatation, especially of the left ventricle.

The main myocardial lesion is hypertrophy, seen on gross and microscopic examination. Other lesions are infrequent and of no great significance.

The irreparable sclerotic lesion is not as a rule favorably influenced by specific treatment. Sudden death is due to acute dilatation or to an anginoid type of failure when the coronaries are involved. When death is delayed it is usually due to failure of the congestive type.

As a rule patients do not survive a two-year period after the diagnosis is made. Exceptional cases may live for many years.

The second main danger of syphilitic supravalvular sclerosis is stenosis and atresia of the coronary arteries. In my series this occurred in about 30 per cent of syphilitic aortitis resulting in death and in about 15 per cent of sudden death due to syphilis of aorta and heart.

The process is always a narrowing, often to complete atresia, of one or both arteries, due to encroachment upon their orifices in the aortic wall by the syphilitic process. Syphilis of the coronaries beyond the aortic wall is rare if we exclude a small amount of adventitial gummatous infiltration which spreads down from the aortic wall and is rarely seen over 1 cm. beyond the orifice of the artery.

If the coronary orifices are congenitally high placed above a base line drawn through the upper attachments
of the aortic cusps, they are much more liable to narrowing than if they arise normally in the sinuses of Valsalva.

The heart is usually normal in size, or occasionally slightly enlarged in the pure forms of this lesion. The muscle is quite normal in appearance and usually shows no evidence of any specific myocardial lesions. Occasionally an inanition atrophy similar to brown atrophy is seen.

Unlike coronary occlusion in arteriosclerosis, in syphilis the process is a very slow and gradual one. Compensatory circulation is often established and keeps the individual alive and often in apparent health for a long time, even with both coronaries completely occluded, when the thebesian circulation supplies the heart muscle.

Myocardial infarction, therefore, practically does not occur in these cases and its absence is almost pathognomonic.

Sudden death is difficult to explain on account of the long duration of the stenosis. Physical strain, fright, emotions, etc., seem to play an important rôle. Syphilitic angina usually occurs in this type of case and not in other forms of cardiac syphilis. It is usually anginoid with atypical symptoms, but sometimes resembles the angina of arteriosclerosis, and even an occlusion in rare instances.

Specific treatment in these cases is usually contraindicated, because of the danger of a Herxheimer reaction or too rapid healing causing a therapeutic paradox.

The third main danger of syphilitic supravalvular sclerosis is aortic aneurysm. In my series, this occurred in 10 per cent of cases of syphilitic aortitis resulting in death but was the most common cause of sudden death, being the direct cause of death in about 37 per cent of medical examiners' cases.

The aneurysm is due to weakening of the aortic wall by the syphilitic, gummatous infiltration. A high maintained diastolic pressure, when the aortic valve is not in-
involved and there is no regurgitation, is usually the important factor in the aneurysmal dilatation.

Cases of long duration are frequently associated with a regurgitation which has been superimposed after the aneurysm has existed for many years.

If there is no regurgitation and the aneurysm is not near enough to the aortic ring to produce dilatation, the heart is usually normal in size, or only moderately hypertrophied.

Death is usually sudden and due to rupture. If the coronaries are involved it may be sudden with, or without, anginoid symptoms. Slow death is due to failure of congestive type, or is a result of pressure on the surrounding structures.

Pain, the main symptom, can often be relieved by proper specific treatment, although the physical signs may increase. Specific treatment is less dangerous and gives better results than in regurgitation or coronary stenosis.

**Other Arterial Lesions**

Of all the arteries, the aortic arch is the chief location for miliary gummatous lesions in late syphilis. The process in the aorta often reaches the great vessels of the aortic arch and produces a narrowing of their orifices.

Syphilis also is fairly frequently seen in the carotid arteries, and carotid aneurysms are by no means rare. This should be expected when we recall the lymphatic drainage from the neck and nasopharynx in syphilis, towards the mediastinum by way of the deep lymphatics. Typical lesions in the femoral arteries have been noted but they are of less frequent occurrence.

It is extremely rare to see a gangrene of the lower extremities which could be definitely laid to syphilitic arteritis. This is accounted for by the fact that the spirochetes from the skin rashes and lesions on the legs drain by way of the superficial lymphatics to the inguinal nodes and away from the peripheral arteries of the legs.
THE PATHOLOGY OF SYPHILIS

Syphilis is frequently seen in the cerebral arteries as a periarteritis, and because of the small caliber of the vessels, an added obliterating endarteritis is liable to produce thrombosis rather than aneurysm.

It may be stated, therefore, that aside from the aortic arch and the cerebral arteries, syphilis plays very little part in lesions of the other arteries.

OTHER INTERSTITIAL AND DEGENERATIVE LESIONS

An interstitial form of pulmonary syphilis, although reported by many clinicians and roentgenologists, is an uncertain diagnosis at autopsy, and as Boyd states, forms one of the most nebulous pathological entities.

Hepatic syphilis in the form of an interstitial cirrhosis is another doubtful pathological entity, the main lesion in the liver being one of multiple large gummas with extensive scarring (hepar lobatum).

An interstitial form of syphilitic orchitis occurs quite frequently, and is much more common than the gumma of the testes. It is often seen at autopsy as the only historical landmark of syphilis and demonstrates the necessity of the routine examination at autopsy of the testes. The testicle on cut section shows pale, grayish, hyaline streaks, sometimes none of the brownish parenchyma being visible, and the tubules string out with difficulty.

Occasionally a splenomegaly is seen in late syphilis, associated with a positive Wassermann and an anemia, the clinical picture being indistinguishable from Banti's disease. In these cases, there is no improvement after antiluetic treatment unless the spleen is removed surgically, the spirochetes seeming to be locked up in the diffuse fibrosis and resisting destruction by treatment. Some authorities have stated that the Banti's syndrome is always syphilitic, but this must be incorrect.

SUMMARY

The development of syphilitic destructive lesions in the nervous and circulatory systems has been given in a
sketchy way outlining certain theories which have appeared to be as near the correct interpretation as we can attain by our present knowledge.

From a clinical and pathological standpoint, I believe that we should regard syphilis of the aorta and heart as an acquired disease (congenital cases being rare) developing insidiously and showing symptoms years after the initial infection.

Death in acquired syphilis of the aorta and heart is almost always due to an aortic regurgitation, or to narrowing or atresia of the coronary arteries, or to the production of an aortic aneurysm, or to any combination of the three great dangers of syphilitic aortitis. Death is due occasionally to unusual lesions caused by syphilis, such as spontaneous rupture of the aorta, dissecting aneurysm, isolated gumma of the heart muscle, miliary aneurysms, etc.

Specific lesions of the myocardium and of the coronary arteries beyond the aortic wall are infrequent, they have been greatly exaggerated, and when they occur are so slight in extent as to be of little practical importance. They rarely embarrass cardiac action, are insignificant in the production of cardiac failure and the slower modes of cardiac death, and are of little importance in explaining sudden death.

The myocardium in syphilis is frequently normal. When the aortic valve is involved, the main myocardial lesion is hypertrophy. Inanition atrophy is occasionally encountered when there is a slow stenosis and atresia of both coronary orifices. Specific lesions of the myocardium are rare, and, when they occur, are slight in extent and of little practical importance.

It is safer and better to assume that the coronaries distal to the aortic wall are usually normal in pure, uncomplicated syphilis, and that coronary occlusions, anemic infarcts, necrosis of heart muscle, replacement fibrosis, aneurysms of ventricular walls, and fibrous myocarditis are almost entirely due to coronary injury dependent upon an
arteriosclerotic process and have nothing to do with syphilis. That rheumatism and other infections produce forms of interstitial myocarditis is obvious. But it appears that syphilis does not play an important rôle in the production of such lesions.

REFERENCES