SYPHILIS OF THE CARDIO-VASCULAR SYSTEM.

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INCIDENCE.

For every patient with syphilitic disease of the cardio-vascular system, there are twenty with heart disease of some other ætiology. It is therefore a much less common cause of heart disease than acute rheumatism and arteriosclerosis, although the latter do not as a rule so quickly cause serious disability. Among the factors that determine whether or not a syphilitic shall develop later cardiac complications are the thoroughness with which the initial infection is treated, the degree of physical stress entailed by the occupation, and the social status of the individual. Men are more likely than women to suffer cardio-vascular sequelæ, particularly in the poorer sections of the community where treatment in the active stages of the infection is likely to be less complete, and in whom laborious and unhygienic conditions of life are likely to prevail. When the cardio-vascular system becomes involved it is usual for the first symptoms to appear about twenty years or more after the primary stage. There are wide variations, but in the great bulk of cases the diagnosis will be made between the ages of forty-five and sixty-five.

Although syphilis accounts for a surprisingly small fraction of unselected heart disease, it is a factor the existence of which it is of the greatest importance to determine. The decision that heart disease is due to chronic lesions in the lungs, or to childhood rheumatism, helps little as far as alleviation of the symptoms or cure of the condition is concerned. On the other hand, the discovery of syphilitic aortitis in a relatively early stage may be of considerable importance to the patient, since it is probable that by active treatment the onset of serious symptoms may be delayed and the expectation of life materially prolonged. At the same time, a syphilitic ætiology may be one of the most difficult to determine. In the first place, the ethical and social factors which are inseparable from infection of this kind frequently render the patient's history wilfully inaccurate. But not only may known syphilitic infection be denied, the patient may have been infected and be yet unaware of it. Needless to say, an admission of gonococcal infection suggests the likelihood of co-existent syphilis. It is obvious that in many cases the conjunction of a history of syphilitic infection twenty years previously, with aortic reflux and a positive Wassermann reaction lead swiftly to an unquestionable diagnosis. On the other hand, with a misleading history, atypical symptoms and signs lacking or inconclusive, the diagnosis may be difficult or impossible.

PATHOLOGY.

In their effect upon the cardio-vascular system the spirochætes and their toxins show a peculiar selectivity. The point of attack is, in almost every case, the arch of the aorta, and practically the whole pathology of cardio-aortic syphilis develops in consequence of the initial gummatous infiltration of the medial coat of the aorta. Thus it is the disruption of the elastic laminæ that allows progressive weakening and dilatation of the vessel, and ultimately in some cases, aneurysm. Again, the extension of the syphilitic infiltration to the commissures of the aortic valve cusps, and later to the valves themselves, leads to aortic regurgitation of variable degree

and occasionally rupture of a cusp; these changes in turn provoke hypertrophy of the left ventricle. But of all extensions of the medial gummatous process the most deadly is that which encroaches upon the orifices of the coronary arteries in the sinuses of Valsalva. This pinching off of the blood supply to the heart is a process that may bring disaster in a remarkably short time, so that a patient may be reduced to a state of acute heart failure within weeks or days of the first appearance of dyspnoea. Syphilitic aortitis has truly been called "a gumma in a dangerous place."

While it is true that the brunt of the damage generally falls upon the aortic arch, infection may produce other results. It may, for example, produce a diffuse syphilitic myocarditis, or local gummatous deposits. Syphilitic mesaortitis may be set up in the coronary arteries and lead to infarction of the heart. But the old conception that syphilis attacks the heart usually in the form of a widespread luetic inflammation of the heart muscle is not supported by the facts of pathology, and in the great proportion of cases such damage as the myocardium suffers in cardio-aortic syphilis is the pure consequence of reduction in arterial irrigation.

The local bulging of the aorta that occurs in the region of the initial aortitis may develop in the course of a year or two into an extensive fusiform dilatation. Sometimes the weakening in the aortic wall is confined to a particular area and then a saccular aneurysm may develop. If the lesion in the aorta is sufficiently remote from the valve ring that the cusps remain unscathed, or if it happens that hypertension co-exists, or if the occupation of the patient entails strenuous exertion, then aneurysm formation is specially likely to occur.

SYMPTOMATOLOGY.

The symptoms of cardio-vascular syphilis are essentially those relating to deficiency in the power of the left ventricle, and to deficiency of the blood supply to the heart muscle. Either dyspnoea or anginal pain is therefore likely to be the presenting symptom. It has been seen that the morbid process attacks the heart at the most vulnerable points, and it might therefore be expected that urgent symptoms would be rapidly produced. Further, the malady commonly ends fatally within two or three years from the time of the first appearance of symptoms. From these considerations it is clear that when a patient first seeks advice his symptoms are likely to be of fairly recent onset—weeks or months rather than years.

Dyspnoea is commonly exertional, it may occur also in paroxysms that awaken the patient at night. Nearly always a patient who has this cardiac asthma admits also that he has been getting short of breath on exertion. The onset of dyspnoea may be extremely sudden, or it may develop and reach a severe grade with orthopnoea in the course of a few weeks or months. Such swift disablement, cardiac in origin, but without obvious cause, the rhythm remaining normal, will suggest the possibility of syphilitic aortitis.

Pain in syphilitic aortitis is likely to be anginal in type, but commonly the relationship to exertion is less concise than in effort angina due to coronary atheroma. It often spreads under the manubrium and up into the throat; while aggravated by exertion it may persist after the cessation of effort. It may be present on waking and gradually pass off in the course of an hour or two. It seems likely that the pain of syphilitic heart disease is a composite one, derived on the one hand from myocardial ischaemia, and on the other from the local lesion in the aorta itself.

A painful attack suggesting coronary thrombosis is unusual in this disease, but when such a seizure occurs at a relatively early age, or in a patient known or thought to be syphilitic, full investigations should be directed to this factor.

Progressive dilatation of the aorta may be the cause of a variety of symptoms. Pressure upon, and paralysis of the left recurrent laryngeal nerve will produce changes in the voice and a failure of normal explosive cough; cough and sputum may arise from pulmonary lesions due to pressure of an aneurysmal sac. Collapse of lung with dyspnoea, or bronchiectasis with characteristic foetid sputum may develop. Girdle pain from pressure on intercostal nerve roots, and the intense boring pain of bone erosion may add to the other discomforts of the patient. The pain of aneurysm in the arch is not infrequently referred to the tip of the right or left shoulder.

PHYSICAL SIGNS.

These will be considered in terms of the various morbid conditions that may be met with in the course of cardio-aortic syphilis. It is clear that there must be a time when the disease in the aorta exists, symptoms and signs yet being absent. Later, there may be symptoms and still no obvious signs. By the time gross signs have appeared, the disease will have passed its early stages.

1. Stage of Aortitis without Aortic Reflux.

Such signs as may be present at this stage will be due to loss of resistance and some enlargement of the affected part of the aortic arch. These changes may cause accentuation of the aortic second sound, together with dullness across the sternum at the level of the second intercostal space, a zone which is in the normal individual resonant to percussion. On viewing the second right intercostal space obliquely in a good light, a pulsation may be seen close to the sternum, this pulsation being also palpable.

It is in this important early stage of syphilitic aortitis that the X-rays may be of supreme value. In the antero-posterior position enlargement of the aorta may be seen, the right border of the ascending part of the arch becoming visible lateral to the superior vena cava, and this part of the right border extending downward to encroach upon the shadow given by the right auricle. In the oblique views it may be found that the aorta is not only enlarged but that the enlargement is local. While high blood pressure or atheroma may cause general enlargement or tortuosity of the aorta, syphilis characteristically affects a certain section of the arch, thereby causing loss of parallelism in the walls of the vessel.

2. Stage of Aortic Regurgitation.

The cardinal signs of aortic regurgitation are a diastolic murmur at the base of the heart, and a collapsing pulse. There are one or two clinical features worth mentioning in relation to these physical signs.

A diastolic murmur at the base means for practical purposes, aortic incompetence. The term "at the base," comprises not only the "aortic area" at the second right intercostal space but also the sternum, at or below this level, and the second and third intercostal spaces on the left side. The murmur may be best heard at any one of these points or even at the fourth left space close to the sternum. In a few cases the diastolic murmur of aortic reflux will be heard only

at the lower end of the sternum, or occasionally only at the apex. In the common event of the murmur being loudest at the base, it may be traced downward along both borders of the sternum or more laterally toward the apex of the heart. When there is a ruptured aortic cusp the vibration in the regurgitant blood may be so gross as to produce a murmur audible not only to the patient but also to the naked ear some distance from the chest; in such cases a diastolic thrill is likely to accompany the murmur. Frequently it is necessary to determine, by timing at the carotid, that a murmur in one phase of the cardiac cycle is diastolic in time. When, as is often the case, to and fro murmurs are heard at the base, timing is unnecessary since one of the two murmurs must be regurgitant.

The term "collapsing pulse" implies one in which the pulsation is abrupt and forcible, and in which the impulse, of short duration, falls away even more abruptly. Such a pulse is due to the vaso-dilatation that, in variable degree, accompanies aortic reflux. Since it is not due directly to reflux it will be no measure of the grade of backflow. Sometimes with an aortic diastolic murmur plainly heard the pulse is not conspicuously, or even at all collapsing, while the blood pressure shows little if any departure from the normal. However, if the pulse is plainly collapsing, free aortic reflux is probably present. A moderately collapsing pulse may be observed also in thyrotoxicosis, advanced arterial disease, and some forms of neuro-circulatory asthenia.

In this stage of syphilitic aortitis there will be, in addition to the signs of aortic reflux, the signs of aortic enlargement already mentioned. The latter signs will be modified in that the aortic second sound will soften or vanish, while radiological examination will reveal the powerful contractions of the left ventricle and the conspicuously jerky and extensive pulsation of the shadow of the aorta as the vessel distends and recoils.

In addition to the signs of first-class importance, namely a basal diastolic murmur and a collapsing pulse, many additional signs due to aortic reflux may be present. An increased pulse-pressure determined by sphygmomanometry, capillary pulsation at the finger nail, and pistol shot systolic sound over the larger arteries may all be observed.

3. Stage of Implication of the Coronary Orifices.

It sometimes happens that the orifices of the coronary arteries become partially obstructed at a relatively early stage in the evolution of aortitis. Symptoms, either dyspnoea or pain, are then likely to develop swiftly, while signs of aortic dilatation or reflux at the valves may or may not be present. The consequences of such obstruction range from sudden death at one end of the scale to diminishing effort tolerance leading to congestive heart failure at the other. The signs of such failure will be identical with those of failure from other causes. Partial obliteration of the coronary orifices frequently produces manifestations of acute insufficiency of the left ventricle—cardiac asthma—which, according to the severity of the condition and the provision of suitable treatment, may or may not lead to acute pulmonary oedema.

When leutic aortitis has brought about coronary obstruction, it is more than probable that changes will have taken place in the aorta or valve cusps so that a careful search for aortic enlargement or regurgitation will almost certainly reveal the basis of cardio-vascular breakdown.

4. Stage of Aneurysmal Dilatation or Sacculation of the Aorta.

The signs of early aortic dilatation have already been mentioned. When such dilatation involves, as it often does, the ascending part of the arch, visible and palpable pulsation may become gradually more and more obvious in the second right intercostal space, and ultimately a pulsatile bulge may appear in this region. At any stage a systolic murmur is likely to be heard over this area, and conducted into the common carotid and subclavian arteries. Careful palpation with the patient in the sitting position and in full expiration will at times also reveal a systolic thrill followed by a diastolic shock produced by the abrupt closure of the aortic cusps. The latter sign will be absent when aortic reflux has been brought about. Aneurysm formation in any part of the aorta is apt to be followed by the formation of a thrombus. The involvement of the ascending part of the arch is thus specially dangerous, not only because aortic reflux is likely to complicate the condition, but also because the coronary orifices may be obstructed by a thrombus.

Aneurysms of the first part of the arch, or of the aorta beyond, sometimes produce no physical signs on examination of the chest, and consequently may be extremely difficult or even impossible to diagnose without radiological examination. Their presence may, however, be inferred from the interference they cause with vital structures in the thorax. When the transverse part of the arch is implicated there may be obstruction of the orifice of one or more great vessels. The pulse will then be noticeably weak or even absent in one or other arm; and the disparity in blood pressure in the two arms will far exceed the 20mm. Hg. that is the limit of the normal. The pulsation in one or other common carotid is then likely to be correspondingly reduced. It should be mentioned that reduction in the radial pulsation alone is not sufficient, and that such reduction must be traced to the brachial and axillary vessels to establish the existence of obstruction at the orifice.

Aneurysm of the transverse part of the arch may cause paralysis of the left vocal cord by interference with the recurrent laryngeal nerve. Since laryngoscopic evidence of this may be present before any symptoms are produced, it is important to make this examination whenever the existence of an aneurysm is suspected. Pressure upon, and damage to the cervical sympathetic trunk will cause inequality of the pupils and perhaps flushing of the left side of the face and neck. Tracheal tugging is a sign commonly sought and infrequently satisfactorily demonstrated.

Radiological examination is always valuable and sometimes indispensable in the diagnosis of aneurysm of the aorta. The shadow of an aneurysm has usually a sharp but not necessarily a smooth outline; subsidiary aneurysms may project from the main mass. Pulsation may or may not be seen on the screen examination of an aneurysm, as a large thrombus may form and partially fill the sac.

DIAGNOSIS.

Whenever symptoms relating to the heart appear for the first time in a man between the ages of 40 and 65, syphilitic aortitis must come into the range of possibilities. Exertional or paroxysmal dyspnoea, or pain of anginal type, of a few months' duration or less, will be suggestive; and specially so if a history of syphilitic or gonococcal infection is given or seriously suspected. Signs of disease in the aorta or regurgitation in the aortic valve in the circumstances outlined above

brings the diagnosis out of the sphere of the possible into the range of the probable. A positive W.R. will be important evidence if the clinical context is appropriate, but it must be remembered that a patient may have this residue of syphilitic infection and yet have cardio-vascular troubles that are not syphilitic. A negative W.R. is obtained in about one fifth of patients with cardio-aortic syphilis; this proportion is less when aneurysm exists. An X-ray screen examination will clinch the diagnosis in a high proportion of cases. Electrocardiography will help little in the diagnosis, although it may give additional information regarding the grade of cardiac damage. Pressure effects such as laryngeal paralysis or boring pain in the chest will sometimes be important clues, while signs of neural syphilis will contribute to the diagnosis in a minority of cases. Lastly, a definite therapeutic effect produced by iodide, mercury or arsenic will point to a syphilitic ætiology.

Difficulty in differential diagnosis will arise at times when there is a history of rheumatic infection and aortic incompetence is found on examination. Obviously, if mitral disease exists also then rheumatic heart disease is undoubtedly present. It should be borne in mind that in only about 5 per cent. of all rheumatic heart disease is the aortic valve involved without the mitral, and that if in a doubtful case there is no evidence of a mitral lesion then the possibility of a syphilitic ætiology becomes much stronger.

A frequent mistake is to regard a systolic murmur and thrill over the aortic area as proving the existence of aortic stenosis. Aortic stenosis is not produced by syphilis; and the presence of such a lesion implies either a rheumatic or atheromatous ætiology. This mistake will not be made if two facts are borne in mind; first, that stenosis implies a rheumatic or atheromatous ætiology, and that rheumatic disease does not involve the wall of the aorta; and secondly, that an aortic thrill and murmur may be as well produced by blood flowing into a dilated aorta as blood flowing out of a stenosed aortic ring into a normal aorta. Consequently, the significance of such thrill and murmur will turn largely upon whether there is evidence of enlargement of the aorta.

At the later ages the differential diagnosis of atheromatous and syphilitic disease of the aorta may be difficult, particularly as these conditions commonly co-exist. Radiological examination frequently is indispensable in solving the problem.

COURSE AND PROGNOSIS.

A diagnosis of cardio-aortic syphilis implies grave heart disease. Many patients survive no longer than three years from the onset of symptoms, while in some the duration is much shorter. Probably the average expectation of life from the first manifestations is in the neighbourhood of five to six years. It is true that in a few instances life may be prolonged without complete incapacity for ten or twelve years, but this is unusual. Probably the thoroughness of treatment of the initial infection influences not only the liability to cardio-vascular disease, but also the course of the malady if it develops.

In the worst event, sudden death from coronary obstruction may occur before the patient has complained of any symptoms. In many cases the patient becomes partially incapacitated by the dyspnoea or pain within a year or two of the onset. The usual termination is by congestive failure due to progressive ischæmia of the

heart muscle. When dilatation of the aorta leads to aneurysm, the prognosis is rather more grave; death may then occur through rupture into the lung, trachea, bronchus, pleura or pericardial sac, or as a result of complications in the lung, chiefly broncho-pneumonia. External rupture of an aneurysm is an event of some rarity.

TREATMENT.

Under this heading must be considered treatment directed to improving the function of the cardio-vascular mechanism, and that designed to combat the underlying infection.

In the first category must be considered the treatment of angina, and of congestive failure. The patient may be seen for the first time suffering from one or other of these dangerous states. If such is the case, or if paroxysmal dyspnoea or increasing exertional dyspnoea are present, then the patient should be put to bed for a period of some weeks. This will relieve effort angina and will often go a long way to abolishing cardiac asthma. Spasmodic anginal pain may persist despite rest, and the usual 1/100 gr. tablets of trinitrin may be employed either prophylactically or as a remedy in attacks. If congestive failure is present digitalis should be employed in the usual way, and restriction of diet, fluids and salt instituted.

When angina or failure have been relieved by suitable measures, or if these symptoms resist treatment ordinarily successful for these conditions when no syphilitic basis exists, iodide and mercury therapy must be started. Potassium iodide, grs. 5-10, may be combined with liq. hydrarg. perchlor. ʒi, thrice daily, or alternatively the iodide may be given separately and the mercury in the form of inunction, a drachm of ung. hydrarg. being rubbed into a different area of skin each day. The more acute the cardiac symptoms, and the more rapid the onset, the more gradually should these medicaments be given.

In no case should arsenic be administered until iodide and mercury have been in use for at least two months; nor should it be given if congestive failure or severe angina is present. Assuming that the foregoing conditions are fulfilled, intravenous injections of N.A.B., starting with gm. 0.15, may be given each week, the dose being increased in the course of five or six weeks to gm. 0.45, and eight injections being given in the course. Such a course may be repeated in four months, and thereafter two courses are generally given each year, starting with gm. 0.3 and increasing to gm. 0.6. During and between the courses of arsenic the use of iodide and mercury will be continued. Antisyphilitic treatment will be continued for the rest of the patient's life.

The treatment of aneurysm is essentially the same as that of the earlier stages of cardio-vascular syphilis, but obviously the results are not likely to be so satisfactory. The pain of aneurysm is not generally relieved by trinitrin, and morphia may be necessary. Large doses of iodide are said to be inadvisable in aneurysm as it is believed that such treatment, by rapidly resolving syphilitic granulation tissue, may remove support and so mechanically cause deterioration rather than improvement.