

his experience the most frequent serious and destructive lesion caused by syphilis is syphilitic aortitis with or without aortic insufficiency or aneurysm formation. It is an accepted fact that the bulb and lower portions of the ascending aorta are the centre of the most striking and characteristic pathological changes in syphilis of the vascular system (Stokes *et al.*, 1946) which take the form of a supravalvular sclerosis (Martland, 1930). Involvement of the more distal part of the aortic arch is commoner in simple arteriosclerosis, while valvular involvement alone is more suggestive of rheumatic or other non-specific cardiac disease. Gordon *et al.* (1942) in a study of 360 cases of chronic syphilitic aortitis noted that atheromatous changes had taken place over the area affected by the syphilitic lesions, and suggest that chronic syphilitic lesions of the aorta predispose to the development of local arteriosclerosis. It would thus appear that, whether or not calcification is present in the aortic valve or in the more distal portions of the aortic arch, calcification of the first part of the ascending aorta is highly suggestive of an underlying or predisposing syphilitic lesion. Thus the radiological sign under discussion appears to have a sound pathological basis.

Summary

The importance of the radiological demonstration of calcification of the ascending portion of the aortic arch as an aid to the diagnosis of chronic syphilis is pointed out. Nineteen cases in which this sign was present are reviewed. The limitations of other diagnostic tests are noted. Of the 19 cases, 7 were serologically negative before treatment and 5 remained positive in spite of treatment.

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**J. S. McCANN AND D. C. PORTER :
SPECIAL PLATE LEGENDS**

- FIG. 1.—Case 4: Left anterior oblique view, showing linear calcification confined to widened ascending portion of aortic arch.
- FIG. 2.—Case 7: Left anterior oblique view, showing calcification involving aortic valve and entire aortic arch.
- FIG. 3.—Case 13: Postero-anterior view, showing calcification in widened ascending portion of aortic arch.
- FIG. 4.—Case 13: Right lateral view, showing calcification in aortic valve and in ascending and transverse portions of arch.
- FIG. 5.—Case 16: Right lateral view, showing extensive calcification confined to widened ascending portion of aortic arch. Note loss of parallelism and traction deviation of oesophagus.
- FIG. 6.—Case 17: Left lateral view, showing calcification in ascending, transverse, and descending portions of aortic arch.

**CARDIOVASCULAR DISEASE IN
SYPHILIS**

A REVIEW OF 1,330 PATIENTS

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The cardiovascular examination of all patients suffering from syphilis has not, so far as we are aware, generally led to that degree of co-operation which prevails between the departments of cardiology and venereology in this hospital. In May, 1945, Sir William Hume undertook the examination of syphilitic patients attending the venereal diseases clinic, and by April, 1947, cardiovascular examination of all such patients had become a routine procedure.

Field of Investigation

Of 1,330 syphilitic patients examined up to December, 1951, 969 were seen prior to antisyphilitic treatment, 79 during the course of treatment but prior to hyperthermy, and 282 after the completion of treatment and surveillance and before being discharged cured.

Method of Cardiovascular Investigation.—The physical examination was generally carried out by the registrar, while screening, which was performed in all but three patients, was done first by Sir William Hume and later by Dr. W. G. A. Swan, so that the radiological opinion was unbiased by the clinical findings. An electrocardiogram was taken on every patient. The function of the cardiovascular department was to assess the cardiovascular status for the venereal diseases department, and antisyphilitic treatment was left in the hands of the latter.

Re-examination.—Patients presenting cardiovascular abnormalities, especially those known or suspected to be of syphilitic origin, were re-examined at approximately annual intervals. With the exception of six who defaulted, all the surviving patients with cardiovascular syphilis were re-examined in 1954. Seventy-three patients out of the whole series died between 1947 and 1954. Fifty died from cardiovascular syphilis and 23 from other causes. In a few cases the necropsy findings did not agree exactly with the clinical diagnosis, and where necropsy evidence was available it was used in the final classification.

Results of Investigation

TABLE I.—Age and Sex Incidence, Together with the Incidence of Cardiovascular Syphilis

Age Groupings	Patients Referred for Cardiovascular Examination				No. Found to have Cardiovascular Syphilis			
	Male	Female	Total	%	Male	Female	Total	%
—29	86	97	183	14	1	1	2	1
30—39	122	153	275	21	6	2	8	4
40—49	154	172	326	24	25	18	43	21
50—59	187	177	364	27	60	29	89	44
60—69	105	52	157	12	42	9	51	25
70+	22	3	25	2	9	0	9	5
Totals	676	654	1,330		143	59	202	
Average age	47	43	45		56	52	55	

The age and sex incidence and the results of the cardiovascular examination are shown in Tables I and II. The high incidence (approximately 35%) of patients subjected to cardiovascular examination under the age of 40 years was solely attributable to the adoption of this procedure for all infected patients, and especially for those who attended for investigation after completing treatment and surveillance for acquired contagious syphilis.

TABLE II.—Results of Cardiovascular Examination

Results of Examination	Male		Female		Total	
	No.	%	No.	%	No.	%
Positive or suspicious cardiovascular syphilis	143	21	59	9	202	15
Non-syphilitic cardiovascular abnormalities	79	12	135	21	214	16
Normal cardiovascular system	454	67	460	70	914	69
Total	676		654		1,330	

The average age of the 914 patients with normal cardiovascular systems was 41 years. In 214 cases (16%) cardiovascular abnormalities were due to causes other than syphilis. The average age of these patients was 51 years and their cardiovascular abnormalities were as follows: hypertension, 151 (71%); rheumatic heart disease, 31 (14%); coronary artery disease, 19 (9%); pulmonary heart disease, 8 (4%); calcific aortic stenosis, 3 (1%); congenital heart disease, 2 (1%). The predominance of females is accounted for by the large number of women with hypertension (96 as against 55 males) and mitral stenosis (27 as against 5 males).

Cardiovascular Abnormalities Due to Syphilis

The final diagnosis in 202 (15%) patients regarded as having cardiovascular syphilis is set out in Table III. Those in whom a constant diagnosis was made (184) have been classed as "positive," and those about whom we are less certain have been classed as "doubtful" (18).

TABLE III

Sex	Total	Positive (184)			Doubtful (18)	
		Uncomplicated Aortitis	Aortic Incompetence	Aneurysm	? Uncomplicated Aortitis	? Syphilitic Heart-block
Male	143 (71%)	18 (12%)	90 (63%)	27 (19%)	8 (6%)	—
Female	59 (29%)	11 (19%)	34 (58%)	4 (7%)	8 (13%)	2 (3%)
Total	202	29 (13%)	124 (62%)	31 (16%)	16 (8%)	2 (1%)

Uncomplicated Aortitis

Uncomplicated aortitis was diagnosed in the presence of dilatation of the aorta to a degree not considered aneurysmal, and in the absence of signs of aortic incompetence or of hypertension. The dilatation was unequivocal in every case so diagnosed, but the diagnosis rests, as it must do, upon the impression formed at fluoroscopy, for, short of angiocardiology, there are no reliable methods for measuring the diameter of any part of the aorta except the isthmus, which is not a region of the first importance in syphilitic aortitis.

The traditional physical signs of aortic dilatation, a basal systolic murmur and a clanging or tambour quality of the aortic second sound, have provided acceptable confirmatory evidence in the presence of radiological signs of a dilated aorta, but they have not been taken as diagnostic if the aorta appeared normal on the screen.

The radiological evidence which we have taken as suggestive of uncomplicated syphilitic aortitis conforms very closely to the standards suggested by Kerley and Shanks (1951). We have looked for a localized or irregular dilatation of the aorta in the absence of a corresponding degree of unfolding or elongation. This change is most often seen in the ascending aorta which bulges to the right of the supracardiac shadow in a rather characteristic way in the postero-anterior view. It is often better seen in the left oblique view, where

the walls of the aorta lose their parallelism and regularity. Calcification of the ascending aorta (Jackman and Lubert, 1945) has proved a useful confirmatory sign on some occasions, but we have not seen it in the absence of dilatation. We have not been able to recognize anything characteristic about the pulsation of the dilated aorta in the absence of aortic incompetence when it is commonly increased.

These changes can be reliably appreciated only at fluoroscopy, and a routine postero-anterior chest film would not be of great value in their detection.

Twenty-nine patients (18 males and 11 females) fell into the group of positive uncomplicated aortitis. It is good evidence of the validity of the criteria we have used, that 15 patients originally diagnosed as cases of uncomplicated aortitis later developed aortic incompetence, and another an aneurysm during the period of observation.

Aortic Incompetence

Aortic incompetence was diagnosed whenever an early diastolic murmur had been heard. This murmur is often faint and must be sought under good conditions in a quiet room with the patient leaning forward and holding the breath in full expiration. It is usually heard better to the left, but occasionally to the right, of the sternum. A diaphragm chest-piece to the stethoscope is useful if the murmur is to be heard when very faint.

We found aortic incompetence in 124 patients (90 males and 34 females). In 16 an early diastolic murmur had appeared during the period of observation, and in these we believe we can date quite closely the onset of aortic incompetence. We have excluded from this group 15 patients in whom aortic incompetence was associated with aneurysm. Where this combination has occurred the aneurysm has been adjudged to be the predominant lesion, and to avoid confusion these patients have been grouped under aneurysm.

Aneurysm

Aneurysm was diagnosed when great dilatation of the aorta was observed at fluoroscopy. It includes the conventional fusiform and saccular types, the latter being much the commoner. An aneurysm has been judged saccular when there was asymmetrical dilatation of the aortic wall and fusiform when a portion of the aorta appeared to be symmetrically dilated in its entire circumference. The division between these categories is easier to make on paper than it is in practice, and it is still harder to say at what point a simple dilatation of the aorta becomes a fusiform aneurysm. We have attempted to divide aortic dilatation into three grades—slight, moderate, and great—and have equated great dilatation of the aorta with fusiform aneurysm. The latter is necessarily large, but a saccular aneurysm can be quite small, as in fact it was in one of our patients.

Aneurysms are quite easily missed on screening, particularly if the patient is obese and the aneurysm small. This, coupled with the difficulty of defining the point at which dilatation becomes an aneurysm, probably accounts for the fact that four of our aneurysms were diagnosed only at necropsy.

The Doubtfuls

This group contained two types of abnormalities. First, there were two patients with heart-block without other evidence of aortic syphilis. Without positive serological evidence, coronary artery disease would be diagnosed, but in these patients a syphilitic origin could not be excluded. Secondly, in the remaining 16 patients (8 males and 8 females) uncomplicated aortitis was suspected but the evidence was insufficient to allow a definite diagnosis. In some the degree of dilatation was too slight to be more than suspicious; in others the dilatation was more definite but there were other factors which could account for it, such as slight hypertension in one and advanced age with marked unfolding in others. The difficulty is naturally greater in the older patients, particularly in those over 65, for the syphilitic aorta is not exempt from the normal processes of ageing, and atherosclerosis and syphilis often coexist.

A few patients have had to remain in the doubtful group because death or default precluded the repeated observation over the years which might have led to a more precise diagnosis.

None of our patients presented pulmonary syphilis.

Clinical Features

The signs and symptoms common to those with aortitis, complicated or otherwise, are listed in Table IV. Among those with uncomplicated aortitis there can be no certainty

TABLE IV

	Uncomplicated Aortitis	Aortic Incompetence	Aneurysm
Total No. of patients	45	124	31
No physical signs	20	—	1
Second sound accentuated	20	21	8
Systolic murmur	15	97	20
Early diastolic murmur	—	124	15
Asymptomatic	36	41	7
Dyspnoea	8	68	21
Angina of effort	2	23	4
Other chest pain	1	26	11
Palpitation	1	19	7

that the symptoms were due to the aortic lesion. Many of these patients were at an age when breathlessness to some degree is very common, and they came from a population in which chronic bronchitis and emphysema are often seen. One of the two patients with angina of effort was proved at necropsy to have coronary atheroma and cardiac infarction—syphilitic aortitis was present, but there was no coronary ostial stenosis. This was the only patient in the group to have an abnormal electrocardiogram. In all these patients the aorta was dilated, and in six the ascending part was calcified.

Aortic dilatation was present in 113 patients with aortic incompetence and in 11 the ascending part was calcified. In 46 patients the left ventricle was normal, in 64 it was moderately enlarged, and in 25 it was greatly enlarged. The electrocardiogram was abnormal in 46 patients.

There were 32 aneurysms in 31 patients; 26 were sacular and 6 fusiform. Seven occurred in the ascending aorta, 7 in the descending aorta, 16 in the arch, and 2 in the innominate artery. Fifteen of these patients had aortic incompetence—there was no association between the site of the aneurysm and the presence of aortic incompetence. In addition to the symptoms already noted, hoarseness occurred in eight, all with aneurysms of the arch, and a brassy cough was found in four. Amongst the physical signs not already listed, abnormal pulsation of the chest wall occurred in 10, tracheal tug in 10, laryngeal palsy in 5, stridor and collapsed lung each in 4, unequal radial pulses in 2, and dilated veins across the chest wall in 2. No patient with aneurysm had an abnormal electrocardiogram in the absence of associated aortic incompetence.

Disability

In an attempt to assess the degree of disability produced by the cardiovascular lesion all patients have been placed in one of four categories corresponding roughly to those suggested by the New York Heart Association (1942). "No disability" indicates that the patient is not limited in any way by his cardiovascular system. "Slight disability" implies that physical activity is a little restricted and that ordinary exertion causes some discomfort. "Moderate disability" implies that activity is much restricted and that less than ordinary degrees of exertion cause some discomfort. "Severe disability" is anything greater than this.

At the initial assessment 87 patients (43%) were without cardiovascular symptoms of any kind and had no disability. They included four-fifths of those with uncomplicated aortitis, a fifth of those with aneurysm, and a third of those with aortic incompetence. In a further 54 disability was

slight, in 47 it was moderate, and in 14 it was severe. All patients in the latter grade had aortic incompetence and 7 were in congestive failure when first seen.

In Table V we have compared our results with eight other surveys dating from 1919. The incidence of aortic incompetence in our series is 9.3%, compared with an average of

TABLE V.—Comparative Incidence of Cardiovascular Lesions among Syphilitics

Author	No. of Cases	Aortic Incompetence	Aneurysm	Uncomplicated Aortitis	Total	Remarks
Hubert (1919)	1,485	3.6%	2.2%	8.8%	14.6%	Aortitis includes myocarditis
Turner (1930)	6,240	2.7%	1.2%	6.2%	10.1%	
Maynard <i>et al.</i> (1935)	346	5%	5%	32%	42%	Aortitis: 19% definite, 13% possible Diagnoses not mutually exclusive
Cole <i>et al.</i> (CCG) (1936)	6,253	4.1%*	1.2%*	4.9%	9.9%	
Cochems and Kemp (1937)	1,000	2%	6.5%	4.2%	12.7%	Aortic incompetence only considered No figure for aortitis given
McDermott <i>et al.</i> (1942)	2,718	3.4%	—	—	—	
Kampmeier (1944)	2,961	3%	1.2%	—	—	Radiological evidence of aortitis not present in all
Dressler and Silverman (1943)	1,270	3%	3.5%	24%	30.5%	
Present series (1955)	1,330	9.3%	2.3%	3.4%	15%	

* Includes all patients presenting a dual picture of aortic incompetence and aneurysm.

3.8% in the other eight groups, in which the incidence is strikingly uniform. The only explanation we can offer for our finding an early diastolic murmur in so many of our patients is that we make a practice of auscultating the chest with the patient leaning forward and the breath held in expiration. We are sure that if this is not done many quiet high-pitched early diastolic murmurs will be missed. The incidence of 2.3% of aneurysms in our series accords reasonably well with the others. When we come to uncomplicated aortitis we are at once struck by the very wide variation in the diagnostic criteria of the various authors. We feel that too much weight has been given to auscultatory findings and antero-posterior chest films; only fluoroscopic examination, short of angiocardiology, can give a satisfactory demonstration of the aorta.

Concomitant Cardiovascular Disease

Concomitant forms of heart disease, actual or potential, were found in 29 (14.3%) of the 202 patients with cardiovascular syphilis (Table VI).

TABLE VI.—Concomitant Cardiovascular Disease

	Male	Female	Total	Aortic Incompetence	Aneurysm	Aortitis Positive	Aortitis Doubtful	Heart block
Hypertension: Systolic over 200	15	13	28	21	4	2	—	1
Diastolic over 100	4	6	10	6	1	1	1	1
Rheumatic: History only	5	5	10	7	1	2	—	—
Signs of mitral disease	1	3	4	4	—	—	—	—
Aortic stenosis	2	2	4	4	—	—	—	—
Cardiac infarction	4	—	4	2	1	1	—	—

Hypertension.—The commonest concomitant cardiovascular disease was hypertension. Of 10 patients with diastolic pressures of more than 100 mm. Hg, 6 had aortic incompetence, 2 had uncomplicated aortitis, one had an aneurysm, and another heart-block.

Rheumatic Heart Disease.—A history of rheumatic fever was given by 10 patients (5 males and 5 females), none of whom had any suggestion of mitral disease. When a syphilitic patient with aortic incompetence gives a history of rheumatism but has no evidence of mitral valve disease, we have considered this to be syphilitic aortic incompetence. None of the four patients with probable concomitant mitral valve disease had a rheumatic history, therefore they are briefly reviewed to justify their inclusion in the series.

(i) A man of 46 with aortic stenosis and incompetence, and calcification and dilatation of the ascending aorta, showed also an apical mid-diastolic murmur, prominence of the pulmonary artery, and slight enlargement of the left auricle. A calcified aortic valve was seen.

(ii) A girl of 18 with congenital syphilis and free aortic incompetence had also an apical mid-diastolic murmur. Fluoroscopy showed a heart with a straight left border and slight enlargement of the left auricle.

(iii) A woman of 51 with aortic incompetence and dilatation of the ascending aorta had also an apical mid-diastolic murmur and definite enlargement of the left auricle.

(iv) A woman of 59 was initially found to have a loud apical systolic murmur, enlargement of the pulmonary artery and of the left auricle, but also well-marked dilatation of the ascending aorta. At a later examination an aortic early diastolic murmur was found.

Aortic Stenosis.—Of 4 patients with aortic stenosis (2 males and 2 females), one had probable mitral disease in addition, and his case has already been considered under rheumatic heart disease. Two others have come to necropsy with evidence of syphilitic aortitis in addition to calcific aortic stenosis, and, encouraged by this necropsy experience, we have hazarded a similar diagnosis in another case.

Cardiac Infarction.—This was diagnosed four times, with necropsy control in two cases. It was not fatal in three and caused sudden death in another, a woman with aortic incompetence and very active aortitis whose left coronary orifice became occluded by a fibrinous plaque shortly after antisiphilitic treatment, in the form of bismuth, had been started. In one of the non-fatal cases the patient died later of intercurrent disease and the cardiac infarction was found to be due to coronary atheroma. This patient had uncomplicated aortitis as his syphilitic lesion. The other three had aortic incompetence complicated in one by an innominate aneurysm.

Syphilis and Cardiovascular Disease

TABLE VII.—Type of Syphilis in Relation to Cardiovascular Diagnosis

Type of Syphilis	Total No. Examined	No Cardiovascular Abnormalities	Cardiovascular Abnormalities	
			Due to Causes Other Than Syphilis	Due to Syphilis
Early contagious ..	163	146 (89%)	16 (10%)	1 (<1%)
Neurological ..	265	148 (56%)	51 (19%)	66 (25%)
Latent, and late clinical (excluding neurological) ..	780	514 (66%)	134 (17%)	132 (17%)
Congenital ..	97	86 (89%)	8 (8%)	3 (3%)
Past history of syphilis*	25	20	5	—
Total ..	1,330	914 (69%)	214 (16%)	202 (15%)

* Clinically and serologically negative, but desired reassurance in view of a past history of syphilis.

The higher percentage of normal cardiovascular systems among patients with early syphilis is to be expected, since the bulk of those patients were below middle age. There was only one patient in this group with aortic incompetence probably due to syphilis. This is likely to be attributable to the efficacy of antisiphilitic treatment in the early stages of the disease, but it might be due to the fact that sufficient time has not elapsed to determine the true incidence of cardiovascular syphilis, even after modern treatment.

Of 265 patients with neurosyphilis, 66 (25%) also had cardiovascular syphilis, while an additional 51 (19%) pre-

sented other cardiovascular abnormalities. The diagnosis of neurosyphilis in those with cardiovascular syphilis was established as follows :

Number with abnormal signs and abnormal cerebrospinal fluid	32
Number with normal signs and abnormal cerebrospinal fluid	24
Number with abnormal signs and normal cerebrospinal fluid	10

The detection of coexisting syphilis in the cardiovascular and central nervous systems is of the greatest importance, and the fact that neurosyphilis was diagnosed exclusively on results of cerebrospinal fluid examination in 24 cases justifies lumbar puncture, where possible, in all patients with cardiovascular syphilis.

Cardiovascular syphilis was found to coexist with ocular syphilis in 15 patients and with tertiary lesions in 11 patients.

Eleven (11%) of those with congenital syphilis had cardiovascular abnormalities, of whom the most important were three with evidence of syphilitic involvement (discussed in detail later).

Serological Findings in Those With Cardiovascular Syphilis

Serological tests for syphilis were performed in 201 patients (one died within 24 hours of his first admission to hospital) : 189 (94%) initially presented a strongly positive serology, 4 (2%) had doubtful serological evidence of the disease, while 8 (4%) were repeatedly negative. In two of the negatives further tests were carried out by Dr. Orpwood Price (Venereal Diseases Reference Laboratory); results were negative in one patient, while in the other a positive treponemal immobilization test was accompanied by confirmatory evidence in three out of the four standard routine tests which he then employed.

Incubation Period and Previous Treatment

Twenty-five (13%) patients gave a definite history of infection from which the incubation period of cardiovascular syphilis could be established, while in 28 (14.5%) the history could only be described as doubtful. With the exception of one patient (Case 1, referred to in detail later), the average incubation period was 29 years, the minimum being 17 and the maximum 44 years.

According to Stokes *et al.* (1934), "the best treatment of cardiovascular syphilis is prophylaxis—that is, at least 20–30 injections of an arsenical with interim heavy metal (bismuth) administered under the continuous system while the patient is in the early stages of syphilis." In none of our patients whose infection could be dated did arsenical treatment in the early stages of the disease approximate to this standard. The minimum was one injection of arsphenamine given to a sailor during the first world war, and the maximum comprised 3.9 g. of neoarsphenamine given to a female patient (Case 1). The latter also received 2 g. of bismuth metal in suspension together with 3.4 mega units of penicillin, but treatment was erratic on account of her frequent default.

Patients Who Merit Special Consideration

Four case histories of unusual interest are discussed in detail below. One patient presented evidence suggesting cardiovascular syphilis four years after reporting with secondary syphilis; the other three have congenital syphilis: two of them present very strong evidence of cardiovascular syphilis, while the third is a possible case of combined rheumatic and syphilitic heart disease. All four patients are alive at the present time.

Case 1.—A woman aged 37 first attended in June, 1946, as a contact of a patient with primary syphilis. She had secondary manifestations of the disease—a generalized maculo-papular rash—and a strongly positive blood Wassermann reaction. She received 3.4 mega units of penicillin, 3.9 g. of neoarsphenamine, and 2 g. of bismuth metal in

suspension, following which she was observed clinically and serologically for four years. Her progress being satisfactory, she was referred for cardiological examination prior to discharge from the venereal diseases clinic in September, 1951, when an early diastolic murmur of aortic incompetence was heard. Fluoroscopy showed no cardiac enlargement and the aorta appeared normal. The electrocardiogram was normal. She has been seen at annual intervals since then—she remains perfectly well, but the clinical findings are unchanged and there has been no alteration in the normal fluoroscopic and electrocardiographic findings. There was no history of rheumatism, chorea, or scarlet fever. This case is quoted to illustrate the difficulty of diagnosis when aortic valve disease is found in a patient four years after treatment for secondary syphilis.

Three Patients with Congenital Syphilis

Case 2.—A woman aged 34 was referred initially from an antenatal clinic on account of doubtful serology. She was found to have Hutchinsonian teeth, a long-standing healed keratitis, and a positive blood Wassermann of low titre. She received only 9.2 g. of arsenic on account of intolerance to this drug, and 12.6 g. of bismuth, followed by 5½ years' of satisfactory surveillance, after which she was referred for cardiological examination. She complained of pain in the upper part of the back, left shoulder, and left arm, which was brought on by exertion and relieved by rest. Her blood pressure fluctuated between 170/90 and 155/75. There was a systolic murmur over the base of the heart and the aortic second sound was accentuated. The first part of the aorta was considered to be dilated and unduly pulsatile on six fluoroscopic examinations between 1950 and 1955. The electrocardiogram was normal apart from one short attack of supraventricular tachycardia. She has improved clinically during the period of observation and now makes no complaint of exertional pain. This patient shows evidence of aortitis which we would accept without serious question in a case of acquired syphilis.

Case 3.—A woman aged 45 had during infancy and adult life received collectively nine courses of bismuth and approximately 7 mega units of penicillin. When we saw her at the age of 45 she had chronic choroiditis, a Charcot hip-joint, and a strongly positive blood Wassermann reaction. There was no history of rheumatism or chorea, but she thought she had had scarlet fever in infancy. She had no symptoms referable to her heart but she was found to have aortic incompetence. Fluoroscopy showed dilatation and calcification of the ascending aorta. The electrocardiogram was normal. There was nothing in clinical or fluoroscopic examination to suggest mitral valve disease. This case gives the strongest possible evidence of syphilitic aortitis with involvement of the aortic valve in a patient with congenital syphilis.

Case 4.—A girl aged 16 was found to have choroiditis of congenitally syphilitic origin according to the ophthalmologist, and for which she had some antisyphilitic treatment during infancy. Both parents were found to have acquired syphilis. She received four courses of nearsphenamine and bismuth, and then, at the age of 18, was seen by the cardiologist. She was found to have free aortic incompetence and a blood pressure of 140/0; a diastolic murmur, present at the apex, was probably accompanied by a mitral opening snap. Fluoroscopy showed a straight left cardiac border and slight enlargement of the left auricle. There was no enlargement of the left ventricle and no dilatation of the aorta. The electrocardiogram had always been normal. The cardiologist follow-up extended over five years, during which time there had been no change in the findings. The patient is of low-grade mentality but has no recollection of rheumatism, chorea, or scarlet fever. This case of congenital syphilis shows a gross lesion of the aortic valve, but has, in addition, evidence of rheumatic mitral valve disease. She cannot, therefore, be regarded as a proved case of cardiovascular syphilis, but, in view of the two cases already quoted, we feel justified in suspecting a double lesion.

Necropsy Findings

Altogether 27 patients with known or suspected cardiovascular syphilis have come to necropsy. The diagnosis of aortic incompetence in 11 instances was confirmed macroscopically, 8 showing narrowing or occlusion of one or both coronary ostia. Naked-eye proof of aortitis was obtained in 12 patients with aneurysm, 4 of whom had aortic incompetence; none showed coronary ostial stenosis. Where sought, histological confirmation was obtained in all (14) cases presenting complicated aortitis. In four patients suspected of having uncomplicated aortitis, naked-eye and histological confirmation was obtained in three, while in the fourth the aorta was reported as normal to the naked eye.

Discussion

Our experience convinces us that the detection of cardiovascular syphilis depends upon careful clinical and fluoroscopic examination of the chest. A simple chest film is not enough to exclude uncomplicated aortitis and therefore fluoroscopy becomes a routine procedure, the oblique positions being essential for the diagnosis of this condition. Repeated re-examination of the patient over a number of years may be necessary before positive evidence of cardiovascular syphilis can be obtained.

The diagnosis of uncomplicated aortitis can be very difficult, especially in older patients or in those with hypertension, a circumstance which may have led us to be too conservative in making a firm diagnosis. In such instances necropsy findings alone can solve the problem. Only seven patients with late syphilis in whom we made normal cardio-aortic findings have come to necropsy—none of them showing evidence of aortitis. On the other hand, in only one patient among four in whom we had suspected aortitis has necropsy proved us wrong.

The high incidence of aortic incompetence in this series (9.3% compared with an average of 3.8% in eight other surveys—see Table V) has surprised us. We have made it a rule never to exclude an early diastolic murmur unless the chest has been auscultated with the patient leaning forward and holding his breath in expiration. We have also been impressed by the superiority of the modern diaphragm type of stethoscope over the older type when it comes to hearing quiet high-pitched early diastolic murmurs. A large number of patients with syphilitic aortic incompetence have no symptoms referable to their hearts. They lead active lives and remain well for years. This has altered our view of the prognosis of this lesion and makes us believe that it is only when symptoms develop and the patients come under observation as cases of heart disease and not as syphilitics that the expectation of life is as short as was once thought.

The establishment of a diagnosis of syphilis in patients presenting a cardio-aortic picture is not always easy. It rests mainly upon serological findings, but evidence of syphilitic manifestations elsewhere in the body may provide confirmatory evidence. Neurological involvement may show itself clinically or may be detectable only by routine examination of the cerebrospinal fluid. A previous history of infection, with or without treatment, may also be of valuable assistance in diagnosis.

Syphilitic involvement of the heart and aorta may be overlooked, especially in its asymptomatic phase and when some other form of the disease dominates the picture.

We are satisfied that close collaboration between syphilologist and cardiologist is necessary for the proper investigation and treatment of syphilitic infection.

Summary

Of 1,330 syphilitic patients referred for cardiovascular investigation, the majority of whom were examined before receiving antisyphilitic therapy, 202 (15%) were found to have cardiovascular syphilis, and 214 (16%) had other non-syphilitic cardiovascular abnormalities.

Cardiovascular syphilis was suspected in one patient who had been treated for secondary syphilis, and in three whose disease was inherited.

Criteria for the establishment of the diagnosis of syphilis are discussed. The great majority of syphilitic patients present undoubted serological evidence of the disease. Time alone will establish the value of the treponemal immobilization test in the remainder.

Difficulties attending the diagnosis of cardiovascular syphilis are briefly mentioned.

Necropsy results in 27 cases confirmed our diagnosis in all but one instance.

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OUTCOME OF UNCOMPLICATED SYPHILITIC AORTITIS

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There have not been many clinical studies of uncomplicated syphilitic aortitis, and on account of the exceptional difficulties of making the diagnosis few are satisfactory.

Some authors, impressed with the frequency of aortitis as a chance finding at necropsy when it had never caused symptoms during life, have claimed that asymptomatic aortitis carries a normal life-span (Heller, 1927; Schlesinger, 1932; Moore, 1941). Among others who have confined their attention to the disease when it is detectable in life opinion is more varied but still comparatively favourable. Moore (1941) suggested that such patients might expect to live for 10 to 20 years after diagnosis. Kalz and Scott (1955) have recently claimed that the expectation of life is never diminished by uncomplicated aortitis, but O'Brien *et al.* (1955) find that it is shortened by at least 10 years.

All the work on this subject has been done in Germany or America, and no one has attempted to study the problem in this country. It therefore seemed of interest to examine the outcome of uncomplicated aortitis in a group of patients attending a British clinic.

Material and Methods

This paper is based upon the experience of 61 patients with uncomplicated syphilitic aortitis seen for the first time before the end of 1951. They were found during the routine cardiological examination of 1,330 syphilitic patients at a special joint clinic established in 1947 at the Newcastle

General Hospital by Professor Sir William Hume in collaboration with Dr. W. V. Macfarlane, Director of the Department for Venereal Diseases, and Dr. W. G. A. Swan, physician in charge of the Cardiovascular Department (Macfarlane, Swan, and Irvine, 1956).

All patients had a history of syphilis, or signs of the disease in other systems, or positive serological evidence in the blood or C.S.F. The disease was of the acquired type in every case save one, in which it was congenital.

All patients were examined clinically, fluoroscopically, and electrocardiographically, and those suspected of having aortitis were re-examined at approximately annual intervals. All living patients in the group were reviewed in 1954 with the exception of two who reported themselves well but refused to attend for examination. No patient was untraced.

Age and Sex.—There were 37 males and 24 females. The ages ranged from 36 to 73, with an average for the whole group of 56 years (males 58, females 53).

Duration of Infection.—Twenty-four of the men were aware of the probable time at which they acquired their syphilis. In these the average duration of infection before the discovery of the aortic lesion was 27 years (13 to 37 years).

Early Treatment.—Ten of the patients had received some treatment at the time of their initial infection. This varied from a single injection of arsenic in one case to 18 months' treatment and surveillance in another. Only two patients seemed to have received more than one course of arsenic.

Diagnostic Criteria

The essential criterion for diagnosis consisted in fluoroscopic evidence of a dilated aorta detected on more than one occasion and in the absence of aortic incompetence. Hypertensive patients were excluded unless the ascending aorta was calcified (two cases) or unless they subsequently developed aortic incompetence, in which case a diagnosis of uncomplicated aortitis was made retrospectively (two cases). The ascending aorta was calcified in seven patients. There is no reliable measurement of the diameter of the aorta apart from angiocardiology, which was not attempted in the present series. The diagnosis rests on a personal impression, formed at fluoroscopy, that the aorta, particularly in its ascending part, is wider than it should be at any given age. Suspicious cases have been re-examined repeatedly, and consistency has been achieved by the fact that all the fluoroscopy has been performed by one observer, Dr. W. G. A. Swan.

Clinical Features

Forty-seven patients were free of any symptoms referable to the cardiovascular system. Among the remaining 14, breathlessness on effort occurred in 11, angina of effort in 4, other sorts of chest pain in 3, and palpitation in 2. There can be no certainty that any of these symptoms were really due to disease of the aorta, for many patients were at an age when some degree of breathlessness is normal, and they all come from a population in which chronic bronchitis and emphysema are extremely common. Moreover, in those with angina it is probable that this symptom was due to concomitant coronary atheroma rather than to coronary ostial stenosis, and this was proved to be so in one case examined at necropsy. No patient complained of paroxysmal dyspnoea.

In 27 patients there were no abnormal physical signs. In 26 there was a basal systolic murmur and in 23 the second sound was increased. In 15 these signs occurred together.

Survival after Diagnosis

In 1954, 51 of the patients were still alive and 10 were dead. The survivors had been under observation for at least three years, extending to 14 years in one patient, who happened to have been thoroughly examined ten years before the inception of the regular clinic for cardiovascular syphilis. The mean observation period was 5.1 years.