CALCIFICATION OF THE AORTA AS AN AID TO THE DIAGNOSIS OF SYPHILIS

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[WITH SPECIAL PLATE]

The limitations of serological tests in the diagnosis of syphilis have long been known, though it is too often forgotten that blood testing may give completely negative results, in spite of an active syphilitic process (Stokes et al., 1946; Wolkin, 1954; Jackman and Lubert, 1945), or, again, that a positive finding may be indicative of a non-specific reaction (Kahn, 1954). The possibility of negative serological reactions, particularly in the old chronic syphilitic, must always be borne in mind, especially in view of the widespread use of modern antibiotics with their ever-present danger of causing a Herxheimer reaction. Nowhere is this more important than in cardiovascular syphilis, where serological positivity is found in only some 70-80% of cases (Stokes et al., 1946). Thorough investigation of the patient must embrace not only a careful case history but also a painstaking clinical examination, together with the help of such diagnostic aids as the electrocardiograph and x rays where necessary. Investigation of the partners, families, and siblings is frequently called for before a complete diagnosis can be made.

Importance of Accurate Diagnosis

The accurate diagnosis of cardiovascular syphilis is of very great importance, not only to prevent a possible Herxheimer reaction as a result of careless therapeutic handling, but also that proper treatment may be carefully and systematically carried out. That such treatment is well worth while has been shown by many workers. Thus Hines and Carr (1930) state that the lives of patients with cardiovascular syphilis are always prolonged by treatment, whilst Moore and Danglade (1930), in a study of 43 patients with aortic aneurysm, have shown that life may be prolonged from an average of 9 months from the onset of symptoms in untreated cases to one of 69 months in adequately treated cases. The same workers have demonstrated, from a study of 90 patients with aortic regurgitation and 8 with various forms of cardiovascular disease, that life may be prolonged from an average of 32 months from the onset of symptoms in untreated cases to an average of 65 months where treatment has been adequate.

A valuable aid to the diagnosis of cardiovascular syphilis has been noted by Schatzki (1942), who pointed out that the presence of calcification of the ascending portion of the aortic arch is highly suggestive of syphilitic aortitis. Wolkin (1954) is also of the opinion that such a finding is a reliable sign of syphilitic aortitis. Jackman and Lubert (1945), who reported 60 radiological examinations in cases of specific aortitis, found linear calcification in the ascending part of the aortic arch in 22.7%, whereas in a control series of 62 cases of arteriosclerosis only 3.2% had calcium in this portion of the arch. According to Thorner and Carter, (1948) calcium was shown radiologically in 39.4% of 38

consecutive cases of syphilitic aortitis. Thus it would appear that the radiological demonstration of calcification in the ascending part of the aorta is relatively common in specific aortitis, while such a finding is seldom made in other conditions (Thorner and Carter, 1948).

Present Investigation

In spite of the references cited above, it seems to us that insufficient attention has been paid to this valuable radiological sign in the diagnosis of syphilis. In the present paper 19 cases of syphilis in which this sign was noted are reviewed. The radiographs reproduced (Special Plate, Figs. 1-6) illustrate various types of calcification found. The outstanding points in these cases are summarized in Tables I and II. It will be seen from Table I that in a number of

TABLE I.—Summary of "Syphilitic History"

Total patients 19	Serological tests:	
Sex M. 13; F. 6	Positive	12
Age limits (years) 51-75	Negative	7
(av. 61)	Cerebrospinal fluid:	
No. with cardiac symptoms 9	Positive	4
Duration of symptoms 4 mths	Negative	8
10 yrs.	Not tested	7
No. with other signs of syphilis 9	No. remaining "Wassermann-	
Approx. duration of infection	fast" in spite of treat-	
where known (years) 20-42	ment	5

TABLE II.—Details with Regard to Calcification of Ascending
Aorta

		Patients
With calcification in ascending aorta		· 19
With calcification confined to ascending aorta		10
With calcification extending into aortic valve		6
With calcification involving more distal segments	of	
aorta in addition to the ascending portion		9
With aneurysmal widening of aorta		4

cases there were no complaints suggestive of a cardiovascular lesion, the signs being, in the main, discovered during the routine medical examination to which all syphilitic patients are subjected in our clinics. Subsequent radiological examination revealed varying degrees of calcification of the aorta (Table II). Symptoms relating to the cardiovascular system included breathlessness on exertion, nocturnal dyspnoea, "tiredness," and anginal syndromes, the breathlessness and anginal syndromes being the most constant. Cardiovascular signs included varying degrees of cardiac enlargement, cardiac irregularities, and murmurs, and were present in most of the cases. In a number of cases there was other evidence of syphilis, such as tabes, Charcot's joint (in one case), and gumma of leg (one case), and one patient with congenital syphilis had rhagades and an old interstitial keratitis of the right eye.

In several cases the radiograph was a valuable aid in establishing a diagnosis on which successful treatment was subsequently based. In at least one case aortic involvement was unsuspected clinically and was discovered only on radiological examination for a chest condition. The limitations of serological testing, whether the result was positive or negative, were again evident, while in at least two of the patients with clinically evident tabes examination of the cerebrospinal fluid was negative. A further point of interest was the percentage of patients whose blood remained "Wassermann-fast" in spite of treatment, another serological limitation which is not infrequently overlooked.

In one case in the present series where a complete family investigation had proved possible, not only was the partner syphilitic, but one of three children—a married daughter—had been congenitally infected.

Discussion

While calcification of the ascending portion of the aortic arch would seem to be a valuable additional aid to the diagnosis of syphilis generally, the importance of syphilitic aortitis per se must be remembered. MacCallum (1940) claims that the most important of all changes produced by syphilis are those which affect the heart and arteries, and in

R. M. MARQUIS: TETRALOGY OF FALLOT

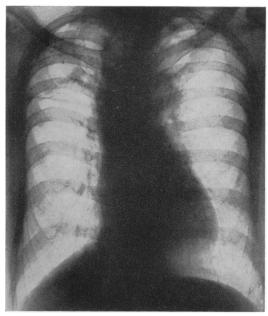


Fig. 3.—Case 2: Anterior radiograph at age 46 showing elevation of apex of heart, prominence of infundibulum, fibrosis of apices of lungs, high position of main pulmonary artery branches, and clarity of lung fields.

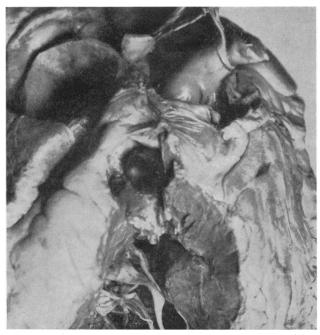
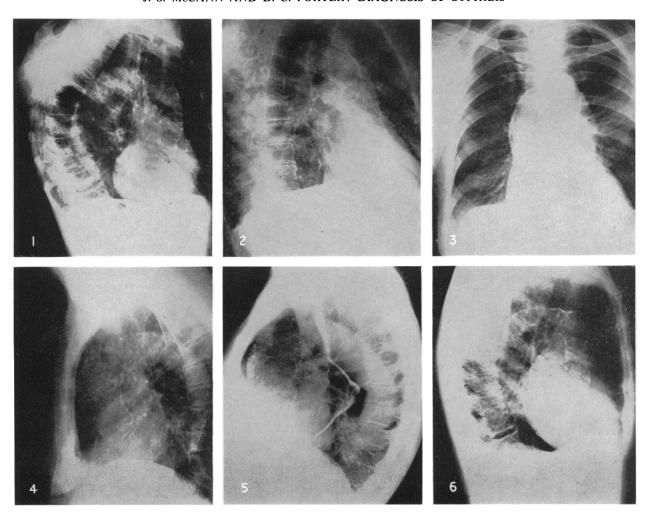


Fig. 4.—Case 2: Anterior view of upper part of right ventricle. Anterior wall cut away to show stenosis of lower bulbar orifice, infundibular chamber, and thickness of right ventricular wall.

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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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REFERENCES

Gordon, W. H. Parker, F., jun., and Weiss, S. (1942). Arch. intern. Med., 70, 396

Hines, L. E., and Carr, J. G. (1930). Amer. Heart J., 6, 142. Halts, E. L., and Cubert M. (1945). Amer. J. Roentgenol., 53, 432.

Kahn, R. L. (1954). Brit. J. vener. Dis., 30, 124.

MacCallum, W. G. (1940). A Textbook of Pathology, pp. 6, 702. Saunders,

Philadelphia and London.

Martland, H. S. (1930). Amer. Heart J., 6, 1.

Moore, J. E., and Danglade, J. H. (1930). Ibid., 6, 148.

Schatzki, R. (1942). New Engl. J. Med., 227, 18.

Stekes, J. H., Beerman, H., and Ingram, N. R., jun. (1946). Modern Clinical Syphilology, 3, 906. Saunders, Philadelphia and London. Thorner, M. C., and Carter, R. A. (1948). Amer. Practit. (Philad.), 2, 301. Wolkin, A. (1954) Radiology, 62, 101.

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Fig. 1.—Case 4: Left anterior oblique view, showing linear calcification confined to widened ascending portion of aortic arch.

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.

-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 With the exception of six who defaulted, all the intervals. surviving patients with cardiovascular syphilis were reexamined 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

	Patients Referred for Cardiovascular Examination			No. Found to have Cardiovascular Syphilis				
	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
6069	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	