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HYPOPLASIA OF THE AORTA

BY

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MALIGNANT AORTITIS:

HYPOPLASIA OF THE AORTA.

THIS condition, in which an aortic lesion comparable to malignant endocarditis presents the clinical features of the latter disease, is very rare. Acute aortitis as the result of various infections—for example, influenza—has long been recognised, but the symptoms are not those of malignant endocarditis and, as Sir Clifford Allbutt[1] has insisted, are prone to assume an anginoid character. Infective or malignant aortitis may accompany and be secondary to malignant endocarditis, the aortic lesion being within reach of vegetations on the aortic valves and merely an extension by contact of the cardiac disease. There is another form of infective aortitis which must be distinguished from that now under discussion; Scheuer[8] has collected seven cases of infective suppuration beginning in the media and adventitia of the aorta, which perforates from without inwards and leads to rupture of the aorta. In Scheuer's own case, which was one of pyæmia, four pints of blood were found in the left pleural cavity. In the case recorded by Vanzetti[12] the cause of the lesion was embolism of the vasa vasorum by the pneumococcus. Infective and ulcerative aortitis, like malignant endocarditis, is more likely to supervene on an old lesion than to occur in a previously healthy part. Thus the aorta is usually already atheromatous, as in F. C. Turner's[10] cases and in Boinet and Romany's[2] case of staphylococcic vegetations in an atheromatous aorta after influenzal bronchopneumonia. As examples of infective aortitis without previous disease of the vessel wall reference may be made to Schweizer's[9] case of a boy,

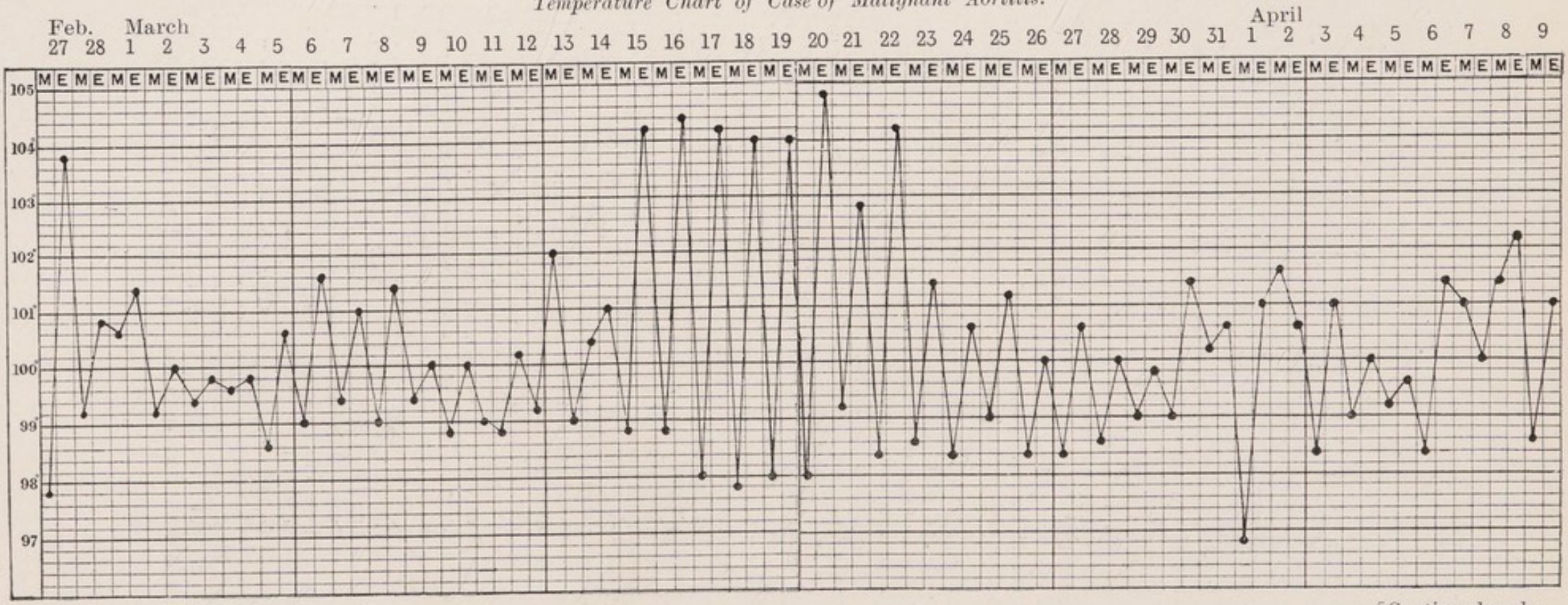
aged 14 years, with septicæmia and pericarditis; the posterior part of the aortic arch was deeply ulcerated and showed adherent clot containing staphylococci. There were multiple abscesses in the spleen and kidneys. Boulay[3] also reported a case of vegetative endarteritis in the thoracic aorta which gave rise to multiple embolisms.

A warrant engineer officer, aged 34 years, was admitted into the Royal Naval Hospital, Haslar, on Feb. 27th, 1915, with fever, gastric irritability, and vomiting. His illness dated from an attack of influenza a month before. On admission he was markedly anæmic without any evidence of ulceration or growth of the alimentary canal, as shown by a negative reaction for occult blood in the fæces, or of intestinal parasites as proved by microscopical examination of the stools. There was no history of syphilitic infection; a Wassermann test was not done. The spleen was never palpable. There was always a trace of albumin in the urine. The blood examinations showed a progressive and grave secondary anæmia with a leucocytosis. On March 1st the red blood count was 2,280,000, the hæmoglobin 45 per cent., and the colour index 0·98. Poikilocytosis and anisocytosis were well marked, and there was some polychromatophilia; no hæmatoblasts were seen. There were 23,000 leucocytes with a differential count of polymorphonuclears 86 per cent., lymphocytes 10 per cent., large mononuclears 3 per cent., and eosinophils 0·5 per cent. On March 12th the red count was 1,950,000, hæmoglobin 40 per cent., and the colour index 1. The changes in the red cells were the same as before. The leucocyte count was 25,800 and the differential count practically identical with that on March 1st. On April 1st the red count was 1,280,000, the hæmoglobin 20 per cent., and the colour index 0·7. The leucocyte count was 19,400. Examination of the blood for malarial parasites, for agglutination of bacillus typhosus, and for micro-organisms by cultivation in bouillon was negative. Except for a minute petechial hæmorrhage near the apex of the heart on May 10th no cutaneous hæmorrhages, rashes, pigmentation, or tender spots were ever seen, and retinal hæmorrhages were not detected. The heart was dilated, and a systolic murmur, loudest over the pulmonary artery, was audible over the whole præcordia, but the character of the murmur did not change while the patient was under observation. The pulse-rate varied considerably; it was almost always over 100, and when the temperature was high ran up to 140. There was pain without obvious swelling or redness in the left shoulder for some days early in April. About April 17th there was thrombosis of the external saphenous vein in the left leg. At first there was no œdema, but as time went on this appeared in the feet and spread to the trunk and scrotum.

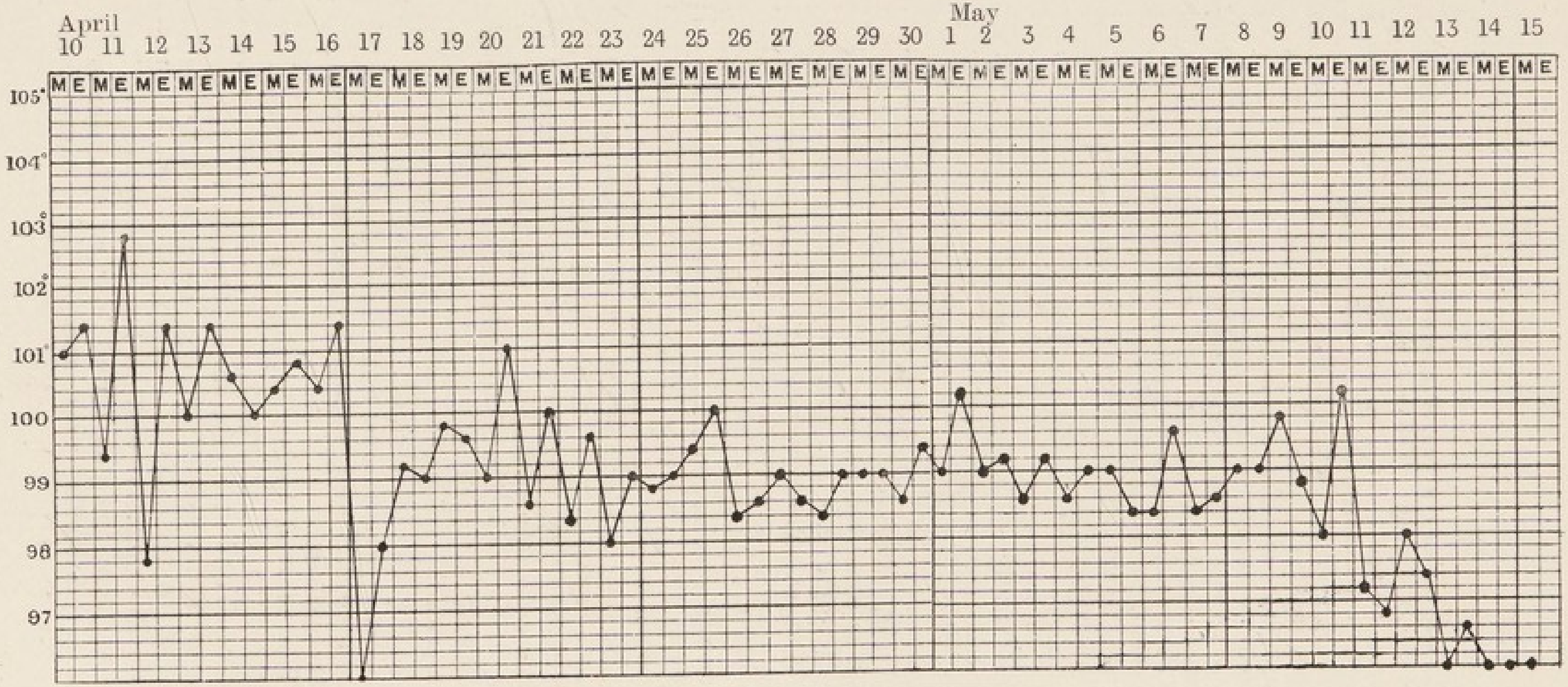
Except for the last few days of life the temperature was always raised (*vide* chart). Directly after admission he had a rigor (temperature 103·8° F.). On March 12th five septic teeth were removed, and from March 15th to 22nd the temperature ran up daily to 104° or above and fell in the morning to normal or below. On March 19th a sensitised streptococcic vaccine (1000 millions) prepared by Dr. M. H. Gordon was given hypodermically and was followed by a rigor. After March 22nd the temperature became lower, and was above 100° until April 16th, from which date it oscillated around 99°. Throughout the illness vomiting was frequent and some epigastric tenderness was commonly present, but no abdominal tumour was ever made out. About April 16th he was very ill with a feeble and rapid pulse (140), and, although he rallied after this for a time, he gradually became weaker and more apathetic, and died quietly on May 16th.

Necropsy.—Both pleuræ contained serous fluid, the right about one pint, the left less. Nothing abnormal was noted about the remains of the thymus. The lungs were normal. The pericardium showed excess of fluid, but was free from pericarditis and petechiæ. The heart weighed 14 oz. The myocardium was pale, but without obvious "tabby-cat" striation. There was not any ante-mortem clot in any of the cavities, nor any recent endocarditis. The aortic valves were much thickened, and two of them were welded together by a calcified bar, but there were no vegetations. In the ascending aorta, 2 centimetres ($\frac{3}{4}$ inch) above the aortic valves, posteriorly and to the left, there was a roughly circular area (A, Fig. 1) 3 centimetres in diameter, or about the size of half-a-crown, resembling the wall of an aneurysm and somewhat bulged outwards. The intima was absent, and thus exposed a slightly roughened surface with undermined, torn and ragged edges, to which two masses of granular fibrin were loosely adherent (B, Fig. 1). It was quite unlike an ordinary atheromatous ulcer. The ulcerated area was so far from the aortic valves that it was clear that it was not due to mechanical damage inflicted by the rigid valves. In removal of the heart this ulcerated area was cut across. The edges have now been sewn together and the specimen (No. 1629) is in the Pathological Museum of the Royal Naval Hospital, Haslar. Deputy Surgeon-General A. G. Wildey, R.N., has most kindly sketched the reconstructed specimen with the omission of the cut which would have confused the photographic appearances. Between its outer surface and some adherent bronchial glands there was a small collection of greenish pus, smears of which did not reveal any micro-organisms. The whole aorta was small, remarkably thin-walled (hypoplasia), and except for a few small patches in the dorsal region very free from atheroma. The abdomen contained some ascitic fluid. The omenta were almost free from fat. A small pedunculated body,

Temperature Chart of Case of Malignant Aortitis.



[*Continued under.*



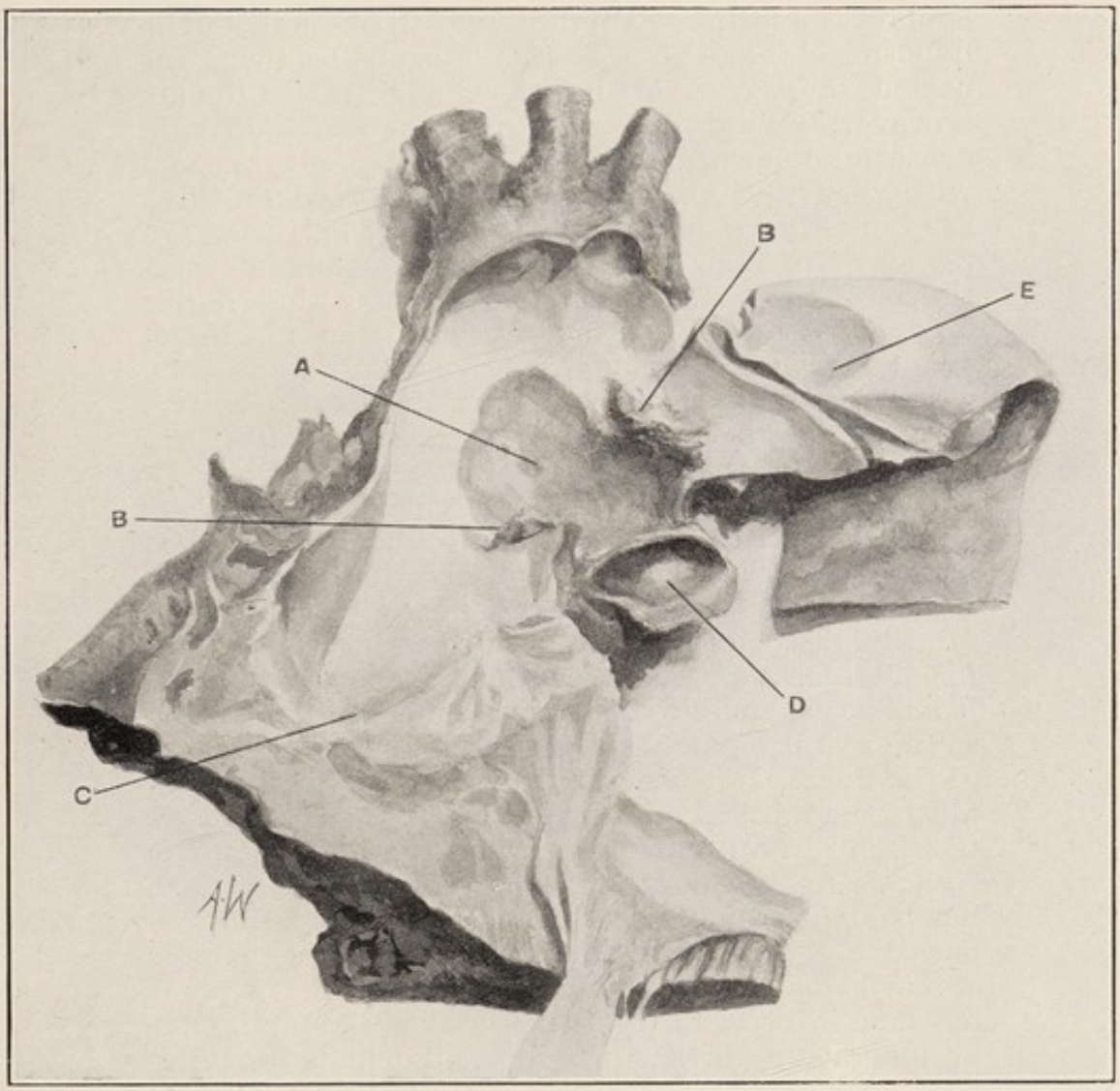
Continued from above.]

the size of half a pea, was attached to the anterior surface of the mesentery. The œsophagus, stomach, and intestines were normal. The spleen, 17 oz., was enlarged and soft, with prominent Malpighian bodies, and contained one large and several small infarcts, white in colour and neither raised nor depressed. There was no softening of these infarcts. The large infarct was firmly adherent to the diaphragm. The splenic vein was normal. The kidneys (right 6 oz., left 10 oz.) were large, pale in colour, and suggested tubal change. Both pelves contained innumerable minute uric acid concretions. I am much indebted to Temporary Surgeon L. Pearce Gould, who carried out the blood examinations and the necropsy.

Remarks.—The most probable diagnosis during life was subacute bacterial endocarditis; but the negative result of the blood culture, which, however, was only carried out once, the absence of splenic enlargement and of obvious infarctions, and to some extent the stationary condition of the cardiac signs, were recognised as militating against this view. The constant albuminuria was, perhaps, not given its proper value as evidence in favour of malignant endocarditis, and the spleen, though obviously enlarged at the necropsy, was never felt during life. An alternative diagnosis—latent carcinoma of the cardiac end of the stomach—was suggested by the association of gastric symptoms, grave secondary anæmia, and fever, while the occurrence of thrombosis was quite compatible with this interpretation. Trousseau, who insisted on the association of thrombosis and gastric carcinoma, exemplified this in his last illness, and remarkable instances of multiple thromboses in this disease have been described by Osler and McCrae.[7] Pernicious anæmia was excluded by the blood count, and especially by the presence of leucocytosis.

The morbid lesion present in this case, which appears to be analogous to malignant or subacute bacterial endocarditis, being situated in the arch of the aorta instead of on the endocardium, was not proved to be infective, as the only blood culture made was negative and no bacteria were seen in a smear from the pus found just outside the aortic lesion. Libman[6] has shown that bacteria-free

FIG. 1.



The aortic region of the heart (C) and the arch of the aorta showing area of malignant aortitis (A) with vegetations (B). The left branch of the pulmonary artery (D) is seen cut across. The commencement of the thoracic aorta (E) is also shown above the line pointing to B. In order to show the thoracic aorta the parts have been somewhat displaced, so that there appears to be a cut between the edge of the ascending part of the aortic arch and the edge of the thoracic aorta. (Drawn by Deputy Surgeon-General A. G. Wildey, R.N.)

periods occur in subacute bacterial endocarditis; but as the blood culture was made on April 4th, when the disease as shown by the temperature chart was active, it would hardly be justifiable to explain the failure to obtain a positive result by the hypothesis that this was an analogous bacteria-free period in a case of infective aortitis. Libman, however, points out that in cases of subacute bacterial endocarditis with high temperatures and with very well-marked oscillations blood cultures are sometimes negative, although bacteria are present in enormous numbers in the vegetations in the heart; and presumably this might hold good in the analogous condition of infective aortitis. Although the view that the case was one of infective aortitis is attractive, it is safer to describe it as one of malignant aortitis.

Hypoplasia of the aorta, described in 1872 by Virchow as an etiological factor in chlorosis, has been somewhat discredited in this connexion, though Burke [4] and van Ritoök [11] have since insisted on this association. Apart from the congenital form, which may be associated with other defects, it has been thought that hypoplasia may be acquired as the result of cachexia, anæmia, and feeble action of the heart. In the majority of cases of aortic hypoplasia the clinical picture is that of a failing heart (Burke). In a number of specimens of true aortic stenosis in St. George's Hospital Museum the aorta has been extremely free from atheroma and comparatively thin. In the present instance aortic stenosis and hypoplasia were associated. A point of some interest in this case is that the malignant or ulcerative aortitis, which is analogous to malignant endocarditis, occurred in a hypoplastic aorta; this sequence of events, therefore, is analogous to the incidence of malignant endocarditis in a damaged or congenitally defective valve. The only focus of infection discovered was the oral sepsis; but according to Burke and van Ritoök hypoplasia of the aorta, like lymphatism with which it may be combined, diminishes the resistance to infection.

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