COARCTATION OF THE AORTA WITH ULCERATIVE AORTITIS.

BY

F. J. POYNTON, M.D., F.R.C.P.

AND

W. P. H. SHELDON, M.D., M.R.C.P.

(From the Hospital for Sick Children, Great Ormond Street, London.)

Although coarctation of the aorta is a rare condition, a considerable number of examples have from time to time been recorded, and the physical signs to which this deformity of the aorta gives rise form a clinical picture which is often definite enough to enable the diagnosis to be established during life.

Just as congenital defects in the formation of the heart predispose it to infection, so may coarctation of the aorta become complicated by the development of a malignant infective aortitis. Four such instances have already been reported, but because of the rarity of the condition, we feel justified in putting on record the unusual clinical signs and the lesions found at autopsy in the following case. We believe this to be the youngest example that has been reported of infective aortitis complicating coarctation of the aorta.

F. D., a boy aged eight years, was admitted to hospital on October 21st, 1927. Ten days before admission he had complained of feeling unwell. Twenty-four hours later he was feverish and became delirious at night. Throughout these days he complained of pain round his heart, in his chest and in both sides of his "stomach." He was breathless, his bowels were loose, and there were some obscure pains referred to the joints without however any evidence of arthritis.

His previous history threw little light on the diagnosis. His mother volunteered that since he was five years of age he had frequently awakened at night complaining of pains in his knees. He had recently fallen and dislocated his elbow, and had taken the anæsthetic badly for the treatment of this injury, and had never seemed well since that date.

On admission he was obviously very ill and lay in a typhoid state, with a temperature of 103·8° F., pulse 140, and respirations 25 to the minute. The heart was not enlarged. The most important physical sign was a systolic murmur audible at the apex beat, in the left axilla and over the back, but with a maximum intensity to the right of the sternum in the third and fourth intercostal spaces, half way between the sternal edge and the right nipple. There were a few rhonchi in the lungs. The abdomen was tender to light palpation; the liver was felt just below the costal arch, and the spleen, which extended an inch below the costal margin, felt firm and was obviously tender.

There were points in the case which suggested an attack of enteric fever of unusual severity, but the positive signs in the heart favoured the view that there was a septic endocarditis. The bacteriological examinations of the urine and the stools by Dr. D. Nabarro threw more light on the case for they excluded typhoid fever, while the blood after 24 hours grew a pure culture of hæmolytic streptococci.

On the day after admission there was noticed a large pulsating external mammary artery the size of a crow-quill, running down vertically from the right axilla. This artery raised the question of coarctation of the aorta, but no other large arteries were detected elsewhere. There was no sufficient reason to believe there was an aneurysmal dilatation of the vessel in the axilla due to a septic arteritis, for the calibre of the artery was uniform and the only unusual feature was its size.

The condition of the boy was quite hopeless; the temperature remained high, incontinence of urine and fæces supervened, Cheyne Stokes respiration and great restlessness developed, and ultimately death occurred seven days after admission.

Post-mortem examination. The body was wasted, and weighed only 38 lbs. Permission to examine the head was refused.

The heart was slightly enlarged, due to some hypertrophy of the left ventricle. The only other abnormality actually in the heart was the aortic valve, which was bicuspid. The innominate artery and the left common carotid artery had a common origin from the arch of the aorta. The ductus arteriosus was closed. About half an inch beyond the origin of the left subclavian artery, the aorta underwent an abrupt constriction, so that the lumen narrowed down to less than half the original diameter. The constriction extended for only a few millimetres, and immediately beyond the constriction there was a patch of ulceration in the tunica intima about as large as a sixpence. The wall of the aorta at this point was weakened, and bulged slightly. The structures in the posterior mediastinum behind the patch of ulceration were normal and not adherent to the aorta. The abdominal aorta seemed of normal calibre.

Apart from some slight engorgement the lungs were normal. The spleen was enlarged and contained several recent infarcts. Both kidneys contained three or four large recent infarcts. The rest of the body appeared to be normal.

On microscopical examination, nothing was found to indicate the site of the constriction in the aorta. In the area of ulceration, the tunica intima was completely destroyed, and the subintima was swollen and lined on its free surface with masses of streptococci. Clumps of organisms extended down into the tunica media, but became less numerous as the deeper layers were approached. In some of the vasa vasorum masses of streptococci could be discerned. Some of the organisms stained poorly, especially those that could be seen lying within leucocytes.

Summary. In brief, the case is that of a boy aged eight years, who died after an acute illness lasting only 17 days. During this period hæmolytic streptococci were isolated from the blood. Autopsy confirmed the suspicion that there was a coarctation of the aorta, and revealed a patch of ulcerative aortitis just beyond the site of the constriction. In addition, the aortic valve was found to be bicuspid, and there was an abnormal arrangement in the origin of the main arteries from the arch of the aorta.

Discussion.

The subject of coarctation of the aorta has recently been reviewed and brought up to date in a very able article by Maude Abbott in Osler's "Modern Medicine."

Bonnett[1] has divided the condition into two groups. In the first, or infantile group, a diffuse narrowing of the arch of the aorta takes place, and terminates at the insertion of the ductus arteriosus, which is usually patent. The process develops during intra-uterine life, and the examples of this group rarely survive infancy. The second, or adult group, to which our case properly belongs, consists of an abrupt constriction of the lumen of the aorta at or about the level of the insertion of the ductus arteriosus. In some cases complete obliteration of the lumen has been found.

There is a striking predominance of the condition in males; in 146 examples of the adult type, Abbott found that 102 were males.

Although grave anomalies in the formation of the heart and large vessels are more frequently associated with the infantile than the adult type, in the latter group certain minor defects are relatively common, and the most usual of these are bicuspid aortic valves and variations in the origin of the great vessels.

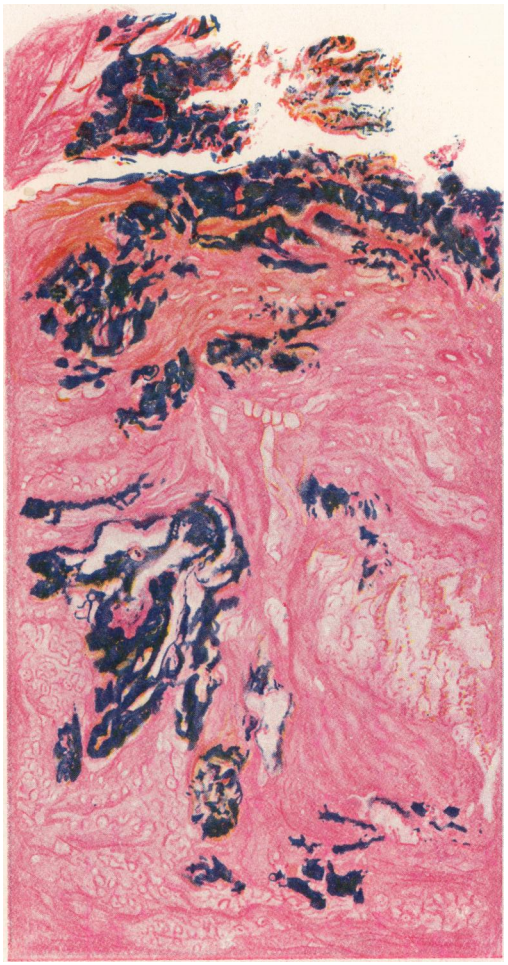

FIG. 2.—Microscopical section of aorta through the edge of the area of ulceration. The ulcer is lined by masses of organisms (stained blue), and these have penetrated into the tunica media, tending to occupy the vascular spaces. Stain: Nile blue and van Gieson. 2/3 objective.

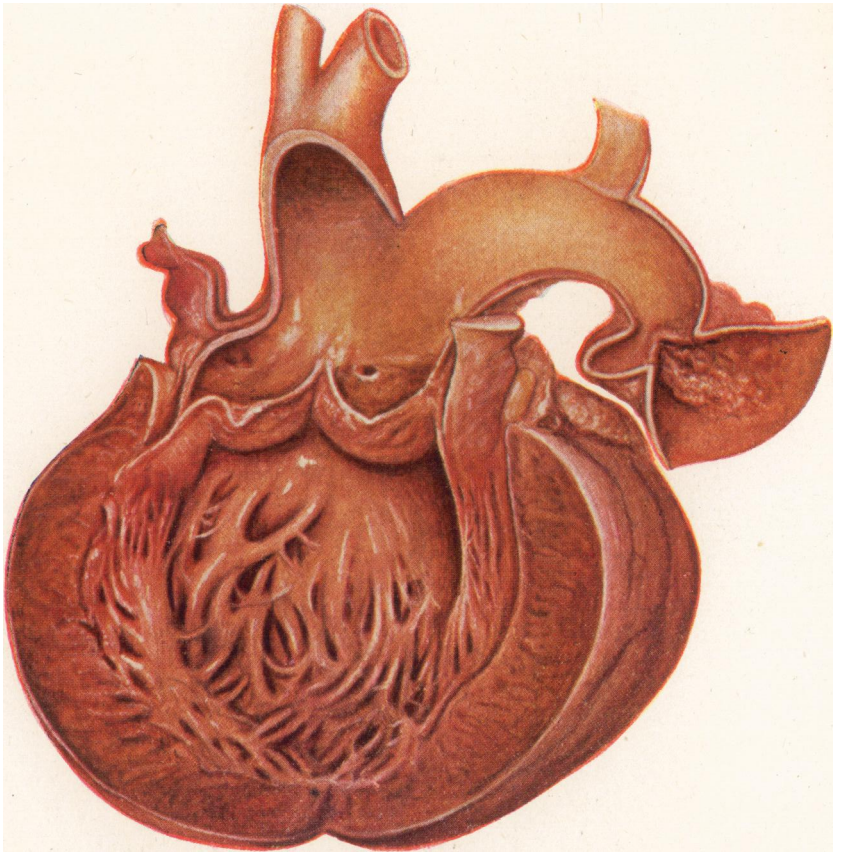

FIG. 1.—Natural size illustration of specimen of coarctation of the aorta. Immediately beyond the point of constriction is an area of ulcerative aortitis. In addition, there is some hypertrophy of the left ventricle, the aortic valve is bicuspid and the innominate and left common carotid arteries arise from the aorta by a common trunk.

Abbott points out the unusual position of the murmurs that have been described and draws attention to the remarkable latency of any symptoms or signs in many cases of coarctation until some disaster, such as has been described here, has supervened. In some cases, presumably those in which a sufficient anastomosis of vessels between the upper and lower portions of the trunk is rapidly and easily formed, coarctation of the aorta may have no effect upon the length of life. The condition has been found at autopsy on a man aged ninety-two. In others, death is brought about through a gradual failure of the left ventricle.

Termination by infective aortitis is quite unusual, and a brief analysis of the recorded cases will therefore be given.

Case 1. (Focken.[2]) Male aged 20 years. Coarctation at the level of the obliterated ductus arteriosus. The aorta just beyond the constriction was the site of ulceration. Vegetations were present here and on the aortic valve, which was bicuspid. Streptococcus viridans was isolated from the blood.

Case 2. (Focken.[2]) Female aged 18 years. Coarctation just beyond the insertion of the obliterated ductus arteriosus. Immediately below the constriction was an aneurysmal dilatation of the aortic wall, due to a small patch of ulcerative aortitis. The aortic valve was bicuspid. A blood culture before death grew hæmolytic streptococci.

Case 3. (Smith and Hausmann.[3]) Male aged 17 years. Coarctation was in this case also associated with a bicuspid aortic valve. There were vegetations on the aortic and mitral valves, and one centimetre below the constriction of the aorta a small saccular aneurysm had formed, and had ruptured into the left pleura. Streptococcus viridans was isolated from the blood.

Case 4. (Reifenstein.[4]) Male aged 10 years. Coarctation of the aorta 2·5 cm. below the origin of the left subclavian artery. Immediately below the constriction a saccular aneurysm had formed, and had burst into the œsophagus. The arteritis was in this case pneumococcal in origin.

An interesting feature of all these cases is that the infection of the aorta occurred at a level just beyond the point of coarctation.

The cause of this deformity of the aorta is unknown. It has been suggested (Skoda) that there is an extension of the tissue of the ductus arteriosus into the wall of the aorta. When the ductus undergoes contraction after birth, the same change takes place in the tissue in the wall of the aorta, and gives rise to coarctation. This view receives some support from the fact that the condition has never been found in the fœtus nor until several weeks after birth, and in addition the ductus arteriosus is found obliterated in over 90 per cent. of cases, whereas in the infantile type the ductus is usually patent. On the other hand the constriction is not necessarily at the level of the entrance of the ductus arteriosus, but, as in our case, may be definitely below this level.

REFERENCES.

1. Bonnet, L. M., *Rév. de Med.*, Paris, 1903, XXIII, 481.
2. Focken, E., *Ztschr. f. klin. Med.*, Berlin, 1924, C, 179.
3. Smith and Hausmann, *Arch. Int. Med.*, Chicago, 1926, XXXVIII, 367.
4. Reifenstein, *Am. J. Med. Sc.*, Philad., 1924, CLXVIII, 381.