A CASE OF COARCTATION OF THE AORTA

BY

A. P. THOMSON, M.D., M.R.C.P., and F. W. M. LAMB, M.D.

(From the Children's Hospital, Birmingham.)

Cases of coarctation of the aorta are sufficiently rare to be worthy of record. No systematic account of the condition is included here as the paper recently published by Poynton and Sheldon¹ mentions the chief references and gives an adequate review of the subject.

In November, 1928, a little girl of five years, the third of a healthy family of 4 children, was sent to one of us (A. P. T.), at the out-patient department of the Children's Hospital, Birmingham, by Dr. J. H. Wright of Four Oaks for an opinion on the significance of a cardiac murmur which he had discovered a few days previously when she had a cold. She was a thin, active and ill-developed child, weighing only 31 lb., the average weight of a girl of three, but nevertheless she had always been regarded by her mother as perfectly healthy, 'like the others,' though admittedly 'on the small side.'

On examination the heart was found to be a little enlarged to the left with a rather diffuse and forcible apex beat in the 5th space, $2\frac{1}{2}$ in. from the midsternal line. A blowing systolic bruit was audible all over the præcordia but most marked at the base, and was conducted up into the vessels of the neck, being loudest over the left common carotid artery. A loud systolic murmur was also heard at the back in the interscapular region, and was there most pronounced in the right paravertebral area at the level of the angle of the scapula. At times the bruit appeared very prolonged, but it was never definitely biphasic; the second heart sound was clearly heard at all areas and was a little accentuated at the base.

A diagnosis of some form of congenital mal-development of the heart, probably of the type of subaortic stenosis, seemed to be satisfactory until a curious difference in the hands was noticed: the left as far as the wrist was uniformly dusky, blue and congested, and presented a striking contrast with the normal right. The difference in the appearance of the hands persisted in all positions and was, in fact, most marked when they were held above the head, for then the right became pallid and no change in the cyanotic condition of the left was observed. The hands and arms were perfectly symmetrical; there was neither œdema nor paresis in the left, and in the absence of any evidence of venous obstruction it seemed probable that the circulatory stasis on the left side was due to some interference with the arterial flow. The fact that the left brachial pulse seemed to be of smaller volume than the right supported this hypothesis and it was possible to hazard the suggestion, which subsequently proved correct, that the child had a coarctation of the aorta extensive enough to involve the origin of the left subclavian artery.

After admission to the hospital for further investigation the results of the examination in the out-patient room were confirmed; in addition, the pulsation in the femoral arteries, though undoubtedly present, was found to be distinctly feeble when compared with that in a child of like development. No sign of a superficial collateral circulation was discovered, but there was no palpable pulsation in the abdominal aorta although the patient was thin and put up with examination placidly. The brachial arteries were very prominent and appeared to be thickened. Mr. E. B. Alabaster examined the retinæ and found the arteries there small and inclined to be tortuous. The appearance of the feet and legs was normal and nothing of note was found in the other systems on physical examination.

An X-ray of the chest by Dr. C. G. Teall showed generalized enlargement of the heart of moderate degree, but no abnormality of the great vessels. An electrocardiograph by Dr. K. D. Wilkinson was normal, apart from slight indications of the changes usually associated with left-sided preponderance.

The most striking new facts elicited by examination in the wards, however, related to the blood pressure. In both arms the tension was found to be much increased, but on the left side it was nearly always slightly higher than on the right. Daily observations at the same hour were made over a period of a fortnight. On the left side the systolic pressures varied from 220 mm. to 200 mm., and the diastolic from 190 mm. to 160 mm., closely following the systolic pressure; the pulse pressure averaged only between 30 and 35 mm. In the right arm the systolic pressure varied between 215 mm. and 175 mm., and the diastolic between 145 mm. and 120 mm., (the high reading was an exceptional one and the diastolic pressure was usually constant at about 130 mm. and did not follow the variations of the systolic pressure as it did on the opposite side); the pulse pressure in the right arm was commonly about 60 mm., roughly double that in the left. In making these observations it was noticed that the 'systolic bang' was much fainter and shorter on the left side. The pressure in the popliteal arteries was found to be fairly constant and equal on the two sides at 125 mm.; it was not possible to determine the diastolic levels in this situation, as no sound could be heard through the stethoscope.

The urine contained a distinct haze of albumen, but the deposit was not abnormal; the blood urea was 49 mgrm. per 100 c.cm. and the non-protein nitrogen was 46 mgrm.

As the blood pressure had been found to be very high in the arms alone and not more than slightly raised in the legs, it appeared unlikely that chronic nephritis could have contributed to the hyperpiesis in any way, but it was thought best to estimate the ability of the kidneys to concentrate urea. The methods of Calvert and Maclean were both employed; with the former the urea concentration seemed decidedly deficient; but with the latter it was found to be unusually good. Three months later after an acute intercurrent illness and shortly before death the Calvert test showed a still more diminished range, although the blood chemistry remained normal and the urine showed only a trace of albumen. In this case, in view of the post-mortem findings, it is clear that this method of estimation of renal efficiency gave a much better result than that of Maclean.

The child was discharged from hospital on November 20th, 1928, and was kept under observation as an out-patient for two months. During this period no material change in her condition was noticed.

On February 1st, 1929, she was re-admitted with lobar pneumonia at the right base. After five critical days she recovered and was about to be discharged early in March, when she developed acute tonsillitis, followed by cervical adenitis from which she rallied very slowly. Swabs from her throat showed no Klebs-Löffler bacilli. Eventually at her parents' request, though still far from well, she was allowed to go home on April 12th, 1929.

During the acute period of her attack of pneumonia several observations on the blood pressure in the brachial arteries were made and it was found that the level had fallen considerably in both, to about 150 mm. systolic and 130 mm. diastolic: curiously enough the constant difference noticed two months earlier (when the pressure in the left arm was consistently higher) did not persist and often a higher pressure was found on the right side. A few days later, however, after the crisis, the blood pressure rose to about 180 mm. systolic and during the week before she finally left hospital it had returned approximately to 200 mm. systolic, and was often above this on both sides: at this time the pressure on the left side was usually distinctly the higher by 10 or 15 mm., as it had been previously.

The marked difference in the appearance of the hands which first drew attention to the child was hardly noticeable while she was acutely ill, possibly on account of general cyanosis, though this explanation seemed rather inadequate at the time. For some weeks before she left the hospital, however, the original condition was restored and she was known in the ward as 'the child with the blue left hand.'

During this prolonged stay in hospital the condition of her brachial arteries and of those in the fundus became worse and clinically they appeared markedly sclerotic: in addition her

mental condition at times was peculiar, and occasionally she vomited. The child, for no apparent reason, suffered from attacks of temper and sometimes from extreme depression: in general she was mentally dull and lethargic.

These symptoms probably were a consequence either of cerebral arteriosclerosis or hypertension, for blood and urine analyses seemed to exclude any possibility of uræmia. Repeated estimations of the blood urea, non-protein nitrogen, calcium and phosphorus were normal, but as already recorded, Calvert's test, which three months previously had given a result indicating renal impairment, now showed a very limited range. The urine remained normal apart from slight albuminuria and was passed in good quantity.

A few days after she left the hospital the child died suddenly at home. Consent to a post-mortem examination was only obtained with difficulty and unfortunately it had to be strictly limited in scope. It was carried out in a small cottage in unsuitable and difficult conditions.

The body was very emaciated (the child had vomited frequently after leaving the hospital) and on opening the thorax the lower lobe of the right lung was found to be firmly adherent to the chest wall: small quantities of fibrino-purulent material were found in the interstices of the adhesions, but there was no empyema. The heart was removed intact with the arch and part of the descending aorta, and one femoral artery and a part of the left brachial artery were dissected out. The abdominal viscera, with the exception of the kidneys, appeared normal. Permission to open the head was refused.

DESCRIPTION OF THE SPECIMENS.

The obvious enlargement of the heart is mainly due to cone-shaped hypertrophy of the left ventricle which forms at least three-quarters of the whole of the anterior surface. So far as could be determined by examination through windows cut in the vessels, there was no abnormality in the arterial valves of the heart, which was not very thoroughly explored in order that its value as a museum specimen might be maintained. The ascending aorta is dilated and has the shiny appearance associated with thickening of the outer fibrous coat: the innominate, left carotid and left subclavian arteries arise in their usual situations from the aortic arch, and the walls of all of them are thickened. Almost opposite to the origin of the left subclavian artery the ductus arteriosus joins the arch; it is less than a $\frac{1}{4}$ -in. in length and is not patent. A little above this point, and for a considerable distance below it, the aorta is greatly constricted but nowhere obliterated: the stenosis is most marked at a point $\frac{3}{8}$ -inch below the left subclavian artery where two small thickened vessels (probably the superior intercostals) take their origin. Below this point the aorta dilates a little, but its appearance is not normal and the total length of the constricted portion exceeds two inches.

Sections from the ascending aorta stained by Mallory's connective tissue stain show an increase in the amount of fibrous tissue, particularly in the region of the vasa vasorum (a region especially affected in syphilitic diseases of the aorta), but the distribution in this case is too diffuse to be suggestive of syphilis. Sections from the stenosed regions show a similar change, but the amount of thickening is greater, and muscular and elastic elements make up the bulk of the increase. Sections of the femoral artery show a surprising degree of fibrosis in both medial and adventitial coats and much of the intima beneath the endothelium shews a fibrous change: muscular elements are sparse, but the evidence of hypertrophy of the muscle is shewn by the prominence of the delicate longitudinally-running muscle fibrils in the intima, probably an early hypertrophic stage of arteriosclerosis. Microscopically the left brachial artery shews considerable hypertrophy of the middle coat, due to increase of the muscular elements, but there is also a patchy fibrosis: here the arteriosclerosis is less well developed than in the femoral artery.

The right kidney was not abnormal in appearance, but microscopically the vessels all show some sclerosis and the glomeruli in some areas are fibrotic. The left kidney was considerably smaller than the right: its capsule was thickened and adherent, and microscopically the secreting elements was found to be disorganized by a diffuse fibrosis: the glomeruli show pericapsular thickenings, adhesions, and hyaline transformation, and many of the tubules are distorted almost beyond recognition into cystic spaces containing hyaline casts. Microscopic cysts are present and the vessels throughout show widespread fibrosis of their coats. The changes are

so advanced in this kidney that they suggest a congenital mal-development of the organ as the basis upon which subsequent changes of vascular, or possibly inflammatory, origin have been superimposed.

DISCUSSION.

The most interesting clinical feature of this remarkable case was the curious cyanosis of the left hand, which in association with cardiac hypertrophy and in the absence of signs of paresis or venous obstruction, first suggested the correct diagnosis. The extremely high pressure in the brachial arteries in contrast with the almost normal level in the femorals furnished indisputable evidence in favour of coarctation.

At the post-mortem it was thought that arteriosclerosis might be found only in the vessels exposed to high pressure, but from the microscopical examination it is clear that this was not the case, for the changes in the femoral artery were more advanced than those in the brachial and there was also considerable sclerosis of the renal and splenic arteries.

As is well known, Bonnet has described two main types of coarctation, the 'adult' and 'infantile.' In the former the stenosis is so limited in extent that the aorta appears to have been ligatured at a point close to the insertion of the ductus arteriosus, which is never patent. Bonnet believes that this type is due to abnormal extension of the tissue of the ductus arteriosus into the wall of the aorta with subsequent contraction as the ductus is obliterated. Clearly our case has no resemblance to this variety.

In the infantile type the stenosis is frequently limited to the so-called isthmus of the aorta (the part that lies between the subclavian artery and the ductus arteriosus which is not much used during intra-uterine life), and Bonnet considers it to be due to failure of development and draws attention to the fact that with it are frequently associated other congenital abnormalities, particularly in the structure of the heart.

In our case the stenosis spreads widely beyond the true isthmus and the only evidence of other congenital abnormality is to be found in the condition of the left kidney, and there not with certainty. Bonnet himself recognized that there were many anomalous cases for which his hypothesis did not account very satisfactorily, and ours appears to be one of them. It may be that the diffuse arteriosclerosis in some way accentuated an original maldevelopment of the isthmus.

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REFERENCE.

1. Poynton, F. J., & Sheldon, W., *Arch. Dis. Child.* Lond., 1928, III, 191.