COARCTATION OF AORTA PRESENTING AS CARDIAC FAILURE IN EARLY INFANCY

BY

J. J. KEMPTON, M.D., M.R.C.P.*Paediatrician, Reading and District Hospitals Group and
Canadian Red Cross War Memorial Hospital, Taplow*

AND

DAVID J. WATERSTON, F.R.C.S.*Surgeon, Hospital for Sick Children, Great Ormond Street,
London*

Of ten cases of cardiac failure in the neonatal period or early infancy from the Thames valley areas served by the Royal Berkshire Hospital, Reading, and the Canadian Hospital, Taplow, and seen by one of us (J.J.K.) during the last three years, four have been cases of coarctation of the aorta. Each, after a period of medical treatment, was transferred to the Hospital for Sick Children, Great Ormond Street, where resection (D.J.W.) was carried out. This paper is an account of the four cases, with a discussion of the problems and the possibilities of diagnosis and treatment which they have seemed to illustrate. No details of investigations are given, and treatment is described in outline only.

Case 1

A baby boy was first seen in August, 1954, in domiciliary consultation, when 12 days old. Difficulty in feeding had begun on the seventh day, the baby having seemed normal until then. The nurse had noticed that he became breathless after sucking for a few minutes, and had choked over several breast feeds. Apart from feeds his mother said that she had twice woken up in the night and heard him panting. He had regained his birth weight of 8 lb. 2 oz. (3,685 g.). There had been no cyanosis, but both mother and nurse thought he looked a little grey sometimes. The family doctor had observed the breathlessness and suspected a heart abnormality.

On examination he did not look ill, and was afebrile. His respiratory rate varied from 30 to 50 and on sucking rose to 90 in about three minutes. His pulse rate was 140 and regular, the arm pulses being easily felt. Heart sounds were normal with no murmurs. Blood pressure in the right arm was 130/70 mm. Hg; femoral pulses could not be felt. Breath sounds were normal. The liver edge was felt at the level of the umbilicus. Limb movements and reflexes were normal and there was no oedema. He was admitted to the Canadian Hospital the following day.

On admission, in addition to the previous signs, some oedema of the legs was noticed, and there were some rales in the lung fields. He was given digoxin, 0.125 mg. eight-hourly, for three doses, then the same dose daily, and was put on salt-free milk feeds. He quickly improved, the signs of failure disappearing within a week. A systolic murmur became audible in the interscapular region at this time. Blood pressure in the arms rose progressively, reaching 180/90 mm. Hg six weeks after admission. Systolic blood-pressure recordings in the legs varied from 40 to 65 mm. Hg. When 6 weeks old a basal systolic murmur became audible anteriorly. He was transferred to Great Ormond Street on October 19. After transfer there was a further rise in arm blood pressure to a maximum of 220 systolic, and signs of failure recurred, needing an increase in the dose of digoxin. Operation was carried out on October 25.

At thoracotomy the coarctation was found to be at the usual site just below the origin of the left subclavian artery. There was a small persistent ductus arteriosus 3 mm. in

diameter just proximal to the coarctation. This was ligated and the narrowed area of the aorta was excised. The aorta was sutured, using 00000 silk thread. The post-operative course was uneventful, and 17 days after operation the baby returned to the Canadian Hospital, where digoxin was gradually withdrawn and feeding with ordinary dried milk established. He was discharged home on November 29 and has been seen at intervals since. At 2½ years he is a healthy-looking child who seems to be developing well and has no symptoms. There is an indistinct basal systolic murmur.

Blood pressure in the right arm remained raised after operation, and in March, 1955, was 130/90 mm. Hg. No pulse could be felt or pressure measured in the left arm. Systolic pressure in the legs was 90 mm. Hg. Now at 2½ years his blood pressure in the right arm is 110/70 mm. Hg. The pulse cannot be felt at the left wrist and systolic pressure in the left arm is 70 mm. Hg. Pressures in the legs are 95 mm. Hg systolic, and femoral pulses are easily felt.

Case 2

A baby girl was first noticed to have noisy breathing and a tendency to choke over feeds in September, 1954, at the age of 4 weeks, and at the time of the mother's first visit to the welfare clinic. On this account she was referred to the family doctor, who in turn sent her to the children's clinic at the Royal Berkshire Hospital. When seen there at the age of 5 weeks she was a healthy-looking infant with no cyanosis or oedema. However, she had gained only 12 oz. (340 g.) in weight from a birth weight of 6 lb. 13 oz. (3,090 g.). Arm pulses were easily felt at a rate of 130. Femoral pulses could be felt with great difficulty. Breath sounds were normal, but the respiratory rate was 40-50 with a little inspiratory indrawing. The respiratory rate rose quickly to about 100 a minute on feeding. A basal systolic murmur was indistinctly audible both anteriorly and in the interscapular region. The liver edge was palpable 2 in. (5 cm.) below the rib margin. No accurate blood-pressure recordings could be obtained in the out-patient department that day, but after admission to the Canadian Hospital the following day systolic pressures were found to be 210 mm. Hg in the arms and 100 in the legs. She was put on salt-free milk and given oral digoxin, 0.125 mg. eight-hourly for five doses and then the same dose daily. Considerable improvement occurred during the next week, feeds being taken with much less respiratory distress and the liver edge becoming less easily felt. There was some weight gain. Further blood-pressure measurements after admission gave figures in the arms of 190-210 mm. Hg systolic, and in the legs 80-90 mm. Hg systolic.

Thirteen days after admission she was transferred to Great Ormond Street. Operation was carried out on December 6. At operation the aortic arch was found to be hypoplastic, and there was a very large pulmonary artery from which a wide persistent ductus arteriosus opened into the aorta just proximal to the coarctation. A palpable thrill was perceptible just above the aortic valve, but pressure recordings revealed no evidence of aortic stenosis. The ductus was divided and the coarctation excised as in the previous case. On releasing the clamps on the aorta the heart rate slowed, with a very marked decrease in cardiac output. Within one minute ventricular contractions had ceased, and in spite of cardiac massage and all the usual resuscitative measures the baby died.

Necropsy showed extensive fibroelastosis affecting the left ventricle and the aortic valve. The right atrium also showed endocardial fibroelastosis.

Case 3

A male infant was sent to the children's clinic at the Royal Berkshire Hospital in August, 1956, at the age of 6 weeks with a letter from the family doctor describing feeding difficulty and failure to thrive. After taking up to 2½ oz. (70 ml.) of a bottle feed he was said to refuse to

suck any more. Birth weight had been 9 lb. 6 oz. (4,250 g.) and at 6 weeks he weighed 10 lb. 3 oz. (4,620 g.). He did not look ill and there was no cyanosis or oedema. Respiration at rest was normal. A feed was watched, and it was seen that he became breathless after two to three minutes. A basal systolic murmur was indistinctly heard in the interscapular region, and the liver edge was palpable at the level of the umbilicus. Breath sounds were normal. Blood pressure in the arms was 160/80 mm. Hg and the femoral pulses could not be felt.

He was admitted to the Royal Berkshire Hospital and given digoxin, 0.125 mg. eight-hourly for four doses and then 0.125 mg. daily. Ordinary dried-milk feeds were given. After digitalization feeds were taken well, and he began to gain weight, reaching 11 lb. 13 oz. (5,360 g.) two weeks after admission. He was transferred to Great Ormond Street, still on his daily dose of digoxin, where, after a period of observation and more investigations, operation was carried out on October 4. At operation there was a small persistent ductus which joined the aorta just proximal to the coarctation. It was ligated and the coarctation excised as in the previous cases. The post-operative course was satisfactory, and after his return home digoxin was gradually withdrawn over a period of two months. He is doing well, though he has had pertussis since the operation. Systolic blood pressures are: arm, 95 mm. Hg; legs, 90 mm. Hg. Femoral pulses are easily felt.

Case 4

This child was born at the Canadian Hospital in September, 1956. He was a large baby with a birth weight of 11 lb. (4,990 g.) and showed signs of cerebral irritation after a difficult forceps delivery. He was kept in the nursery for five days and was at first tube-fed. He was thought to be well and had regained his birth weight on discharge on the eleventh day. Two weeks later his mother took him to see the sister in the obstetric nursery; lactation had failed and bottle feeds were not being taken well. The sister watched a feed and saw that the baby became breathless and had a choking fit after sucking for a few minutes, and so kept mother and baby until the paediatric round later in the day.

On examination then the baby had a good colour and was not breathless at rest. There was no oedema or cyanosis. The liver edge was palpable below the level of the umbilicus. Breath sounds were normal. A systolic murmur faintly audible over the cardiac base was more easily heard between the scapulae. Femoral pulses could not be felt. Blood pressures by the flush method were: left arm 160 mm. Hg, left leg 70 mm. Hg. He was admitted and given digoxin, 0.125 mg. eight-hourly for four doses and then the same dose daily. Nine days later, when signs of failure had cleared, he was transferred to Great Ormond Street, still on a dose of 0.125 mg. of digoxin daily, and taking feeds of an ordinary dried-milk mixture fairly well. However, after admission he again showed signs of failure which did not respond satisfactorily to treatment, and urgent operation was decided upon.

Operation was planned for November 1, but fairly severe pulmonary oedema developed during induction of anaesthesia; operation was deferred and a further attempt was made on November 8. On this occasion cardiac arrest occurred immediately following tracheal intubation. An urgent thoracotomy was performed and the heart restarted after cardiac massage. Further procedures were not attempted at this time, but on November 19 the coarctation was successfully resected after ligating a large persistent ductus arteriosus which entered the aorta proximal to the stenosis. Post-operative progress was uneventful.

After return home digoxin was gradually withdrawn over a period of three weeks. The baby appears to be thriving. Blood pressures four months after operation were: arms 130/85 mm. Hg, legs 85 mm. Hg systolic. Femoral pulses were easily felt.

Discussion

It has been established that coarctation of the aorta is a common cause of cardiac failure in early infancy (Keith, 1956). The incidence of failure itself in babies is hard to assess. The essential early symptoms—breathlessness on sucking, and subsequent choking attacks as the respiratory rate rises—are very likely to be disregarded, and an experienced doctor or nurse may regard the picture as that of mechanical feeding difficulty unless a feed is watched with the chest exposed. The essential physical sign is liver enlargement, and palpation of the liver edge at this age, even when there is much enlargement, is of the same order of difficulty as palpation of the tumour of pyloric stenosis, and requires the same technique and patience. There may be no oedema, no cyanosis, and no chest signs. No murmur may be audible in the early weeks.

Inhalation of milk and gastric contents, together perhaps with early pulmonary oedema, may lead quickly to a picture of respiratory distress with distended chest, costal indrawing, and downward displacement of the liver edge in which the cardiac origin of the illness may be very easily overlooked—may not, indeed, be at all apparent—so that cases are even more likely to be missed if they reach this stage.

Once the basic diagnosis of failure has been made, the diagnosis of coarctation will of course depend on observation of limb pulses and blood-pressure measurements; the latter, whether by auscultation (for which a number of small-sized cuffs are needed) or by the flush method, needs time and a degree of practised skill, and there is a considerable personal error.

Final decision on the type of coarctation present, and the indication for immediate or late operation, will need more detailed investigation in a special centre.

Bonnet (1903) divided coarctation into an infantile developmental type, with hypoplasia of the arch and narrowing occurring before the opening of a persistent ductus arteriosus, and an adult type in which the fibroelastotic process closing the ductus was assumed to have extended into the aortic wall, producing a constriction. This division was accepted by Maude Abbott (1928) and is still sometimes used, though the two separate processes of development are not accepted, and it is well known that the infantile form may be found in adults, and vice versa. Mustard *et al.* (1955), writing with special reference to the first year of life, divided their cases into pre-ductal and post-ductal types, the first approximating to Bonnet's infantile type and having a very high mortality. Previously Johnson *et al.* (1951) had devised a classification based on the state of the ductus arteriosus and its relationship to the stenosis and on the amount of collateral circulation.

It is perhaps best, considering the almost infinite possibilities of abnormality in development from the 12 embryonic arches, not to attempt any exact classification. We can be sure that no two cases will be exactly alike.

Resection of coarctation was first carried out in young adults by Crafoord and Nylin (1945) and Gross (1945, 1950), and there are now a number of published series of successfully treated cases; the subject has been reviewed recently by Campbell and Baylis (1956) and also by Cleland *et al.* (1956), who surveyed 52 cases in patients aged 1 to 55 years, 40 of whom had undergone resection.

Calodney and Carson (1950) reviewed 22 cases diagnosed in infancy, all the patients dying in cardiac failure, and suggested that in some cases operation might be possible. A case successfully undergoing resection at 2 weeks was recorded by Baronofsky and Adams (1954). Mustard *et al.* (1955), of Toronto, described 15 cases operated upon in infancy, the youngest at 11 days. There were five deaths, all cases of "infantile" or pre-ductal coarctation, the remainder having survived and continuing to do well. Nouaille *et al.* (1955) described 15 cases in early infancy, five of which successfully underwent operation. Of 10 patients not operated upon, eight died: but one of the two survivors lost all symptoms after a period of severe failure.

Gross (1953) stated that the majority of these babies can be tided over their period of embarrassment by medical means, and that the best chance of successful operation will come later, between the ages of 8 and 12 years.

Lang and Nadas (1956), reporting nine cases treated medically, also suggested that most of these infants can be managed satisfactorily by medical means, but added that surgery should be performed early if medical management fails.

Though no classification of cases of coarctation is really satisfactory a certain therapeutic grouping appears reasonable. First there will be cases where there is gross abnormality of the arch, often with other anomalies, sometimes with very extensive endocardial fibroelastosis, and in which the deformity is such as to be incompatible with survival. These patients are likely to die within the first few hours or days. A second group will comprise those who have early and severe cardiac failure which proves difficult to control, and who will need surgery as soon as possible if they are to survive. Thirdly, there are those whose symptoms are less severe and more easily managed, and who have a tendency to marked spontaneous improvement after the first few months. Some of these might well be kept under supervision and operated upon later, after the general improvement has occurred and all signs of failure have disappeared. Finally, there is a fourth group of cases in which the early symptoms are slight or absent, and which are unlikely to be recognized in infancy. Retrospective inquiry into feeding difficulties, choking attacks, and breathlessness in the infancy of adults found to have coarctation would be of doubtful value, and, so far as we are aware, has never been undertaken. This group therefore includes all adult cases.

Until recent years this condition in infancy was not thought to be important; if it was of the infantile type death was inevitable, and if of the adult type no symptoms would occur. Reports of larger series, and the individual study of our four cases, would appear to show the importance of coarctation in babies, and our cases seem specially to show the way in which the successful management of this not uncommon condition depends on care at all levels. Cardiac failure in babies presents as feeding difficulty, often developing quickly, after choking attacks, into a cardio-respiratory illness in which the diagnosis becomes very much more difficult and the prognosis very much worse. Early diagnosis before this occurs can result from alertness to the possibility on the part of the family doctor, the midwife, and the nurse. Confirmation of diagnosis and immediate investigation and treatment can be carried out in the area paediatric department. This will consist essentially in really close ward observation and management of feeding, in x-ray screening, and, generally, in digitalization with serial electrocardiograms. Salt-free milk powder will be helpful in some cases. This period of emergency care is followed by more investigation and more detailed assessment in the specialized unit in which resection may be carried out, either at once or later. Later, care of those operated upon and of those in whom operation is deferred after assessment can again be undertaken by the paediatric department and family doctor, and will usually include the gradual weaning off digitalis. Blood pressures may in some cases not return to normal figures for several months after operation, so that continued recordings are advisable.

There is the possibility, mentioned by Gross (1953) and others, that stenosis may occur at the site of resection. Animal experiments, on the other hand, have suggested that growth of the scar may occur. Only time will show. Of more immediate importance is the inescapable impression that many of these babies must die as cases of bronchopneumonia, the real cause of their illness remaining unrecognized.

Summary

An account is given of four cases of coarctation of the aorta presenting as cardiac failure in early infancy. In each case resection was carried out after medical

treatment. There was one death; the remaining three children have done well. The history and presenting features in each case are described in detail in order to show the importance of the recognition of early symptoms and signs in those babies who may appear to be cases of mechanical feeding difficulty or chest infection.

The management of these and similar cases is presented as a combined responsibility.

We wish to acknowledge the part taken by Dr. R. E. Bonham Carter in the care of these cases, and to thank him for much help. These four babies are to be included in a more extensive review of congenital heart disease in infancy, from Great Ormond Street, which is in course of preparation.

REFERENCES

- Abbott, M. E. (1928). *Amer. Heart J.*, 3, 574.
 Baronofsky, I. D., and Adams, P. (1954). *Ann. Surg.*, 139, 494.
 Bonnet, L. M. (1903). *Rev. Médecine*, 23, 108.
 Calodney, M. M., and Carson, M. J. (1950). *J. Pediat.*, 37, 46.
 Campbell, Sir M., and Baylis, J. H. (1956). *Brit. Heart J.*, 18, 475.
 Cleland, W. P., Counihan, T. P., Goodwin, J. F., and Steiner, R. E. (1956). *British Medical Journal*, 2, 379.
 Crafoord, C., and Nylin, G. (1945). *J. thorac. Surg.*, 14, 347.
 Gross, R. E. (1945). *Surgery*, 18, 673.
 — (1950). *Circulation*, 1, 41.
 — (1953). *Ibid.*, 7, 757.
 Johnson, A. L., Ferencz, C., Wigglesworth, F. W., and McRae, D. L. (1951). *Ibid.*, 4, 242.
 Keith, J. D. (1956). *Pediatrics*, 18, 491.
 Lang, H. T., and Nadas, A. S. (1956). *Ibid.*, 17, 45.
 Mustard, W. T., Rowe, R. D., Keith, J. D., and Sirek, A. (1955). *Ann. Surg.*, 141, 429.
 Nouaille, J., Mathey, J., Schweisguth, O., Labesse, J., and Binet, J. P. (1954). *Sem. Hôp. Paris*, 30, 2610.

CAT-SCRATCH DISEASE

BY

E. HINDEN, M.D., M.R.C.P.

Paediatrician, Whipps Cross Hospital

Cat-scratch disease is an illness which is not well known in this country. It usually presents as a mass of inflamed lymph nodes in the groin, axilla, or neck, which commonly proceeds to suppuration, though occasionally it may resolve. Daniels and MacMurray (1954) present a comprehensive account of the disease, based on 160 cases (including 27 personally observed); Brand and Finkel (1956) describe three cases and give a bibliography of all the others reported to date in this country. The former authors write: "Cat-scratch disease may simulate a wide variety of lymph-node lesions such as tularaemia, infectious mononucleosis, lymphosarcoma, Hodgkin's disease, tuberculous adenitis, lymphogranuloma venereum, and both benign and malignant disease." The following cases illustrate some of these diagnostic pitfalls, showing the need to consider all data, clinical and pathological, in order to reach a correct diagnosis. They also include an unusual familial relationship.

Case 1

A boy aged 2 years 2 months was admitted to hospital on September 1, 1954, from the surgical out-patient department. He had been sent there by his family doctor because of a lump which had appeared in his right groin five days previously. It was neither painful nor tender; it was hard and irreducible; there was no impulse on coughing or disturbance in his general health. It was thought that it might be a hernia with impacted omentum.

At operation later that day the mass was found to consist of enlarged lymph nodes, and it was excised. The histological report on the specimen stated: "Marked reticulo-endothelial proliferation in lymph nodes with a few mitotic figures and a suggestion of granulomatous pattern. I cannot