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COARCTATION OF THE AORTA AND PREGNANCY

REPORT OF FIVE CASES

BY

LEONARD ROSENTHAL, M.D.

Physician, Geriatric Unit, Pontefract and Wakefield Hospital Groups; late Physician-Superintendent, Wakefield General Hospital

Coarctation of the aorta is an uncommon finding in adults and shows its higher incidence in males in a proportion of 3 males to 1 female (Abbott, 1928). The association of this anomaly with pregnancy must therefore be of some rarity. Benham (1949) was able to find records of only 53 cases in the literature and added three personal reports. To these can be added the following cases: Dieckmann (1941), 4; Magnin (1944–5), 1; Novak (1947), 1; Clark and Koenig (1947), 1; Cohen (1949), 1; Marcorelles (1949), 1; Money (1949), 1; Sundfør (1950), 3; Vander Veer and Kuo (1950), 2; Brown (1950), 4; Greig and Parker (1951), 1; Bond (1951), 1; Kerr and Sodeman (1951), 5; Soulié *et al.* (1951), 3; Notter (1952), 1; Miller and Falor (1952), 2; Stéphan (1952), 1; Peterson *et al.* (1953), 1; and Taylor and Pollock (1953), 1; making a total of 91 recorded cases.

In view of the infrequency of pregnancy complicating coarctation of the aorta the assessment of the risks involved and the prognosis and management of any individual patient become considerations of great importance and difficulty for the physician and obstetrician whose experience of this association of conditions must necessarily be limited. It would seem desirable that as many records as possible be reported so that a firmer basis for judgment may be provided. With this in view the following five cases are reported.

Case 1

The patient, aged 25, was referred from the local antenatal clinic at the third month of her first pregnancy for the diagnosis of a suspected heart lesion. A "rheumatic heart" was said to have been discovered when she was 5 years old, and from about the age of 10 she had had symptoms very suggestive of mild intermittent claudication. These lasted on and off for about five years. The only symptom of note after this had been mild dyspnoea on exertion, which she thought was probably a little more prominent since the beginning of her pregnancy. Except for this and very occasional "bilious attacks" (vomiting, mild headaches, but no eye symptoms) she was perfectly fit and well.

Examination showed the classical signs of coarctation of the aorta. Blood-pressure readings were: right arm, 170/100; left arm, 150/95; right leg, 120/100; left leg, 115/90. The heart size was within normal limits. She had a mild ichthyosis, mostly involving the skin of the trunk, upper arms, and thighs, and this had been present since childhood. She had no other abnormal physical signs. *X*-ray examination of the chest showed a poorly developed aortic knuckle and notching of the third, fourth, sixth, seventh, eighth, and ninth ribs of both sides. Blood counts, blood urea, and Wassermann reaction were normal. Intravenous pyelography studies and an electrocardiogram were within normal limits. The amylobarbitone test resulted in an average blood pressure of 130/95 compared with 155/100 before the test.

She attended the medical clinic regularly and the pregnancy proceeded uneventfully. At the 27th week a slight generalized enlargement of the thyroid gland was noted. She was admitted to hospital at the 36th week for a final rest prior to confinement. One week later she began to complain of nausea, hot flushes, palpitations, and precordial pain. Over the course of three hours her pulse rate rose from 80 to 115 a minute, the systolic blood pressure from 135 to 165 mm. Hg, and the diastolic pressure from 95 to 100 mm. Hg. It was decided to perform caesarean section at once. The lower segment operation was done under spinal analgesia, and a living infant weighing 5 lb. 6 oz. (2.4 kg.) delivered. The patient was not in labour and there was much bleeding from the thickened lower segment of the uterus. It was noted that the abdominal aorta was half normal size. The puerperium was uneventful, and she was discharged from hospital after three weeks.

The patient was seen again at the medical clinic three months later. She had remained fit and well with no complaints except for slight dyspnoea on exertion, which was no worse than before the pregnancy. There were no notable changes in the physical signs. She remained in good health for five months after delivery. One night, about one and a half hours after going to bed, she awoke, told her husband she felt "queer," rapidly became unconscious, and died within half an hour. A post-mortem examination was not carried out.

Case 2

A primipara aged 21 was referred from the local antenatal clinic for diagnosis of a suspected heart lesion. She had had no serious illnesses as a child and had played games and cycled as other girls of her age. She had no complaints of headaches or pains in the legs. Eighteen months previously she had been off work for three months with dyspnoea on exertion, lethargy, and ankle oedema. These symptoms returned when she became pregnant, but on stopping work she regained her former good health.

On examination she was found to be 20 weeks pregnant. The blood pressure in the arms was 165/85 mm. Hg, and the systolic pressure in the legs, by palpation of the dorsalis pedis artery, 105 mm. Hg. A systolic murmur could be heard at the base of the heart at the level of the second rib on the left side close to the sternum. A loud systolic murmur was heard over the whole posterior chest, maximal in the mid-zone on both sides. A systolic thrill was felt in the left supraclavicular region. *X*-ray examination showed the heart to be normal in size and configuration, though the aortic knuckle was not seen. Rib notching was not present.

The pregnancy continued uneventfully until the thirty-eighth week, when she was admitted with uterine haemorrhage. A central placenta praevia was diagnosed and a living infant of 5 lb. 14 oz. (2.7 kg.) delivered by lower segment caesarean section. No note was made of the size and pulsation of the abdominal aorta. Except for a mild pyrexia and anaemia the puerperium was normal. On the tenth day the cardiac bruits were unchanged. No collateral circulation could be felt or seen. The blood pressure in the arms was 150/75 mm. Hg, while that in the legs could not be determined. The femoral artery pulsations were not felt.

Case 3

A woman aged 43 was referred to the local maternity hospital because of a raised blood pressure, upper abdominal pain, and malaise three weeks before her fifth child was due. She had had four previous normal pregnancies and labours, and her past medical history was uneventful.

On examination, except for a blood pressure of 130/90 mm. Hg, no abnormalities were noted. The urine was free from albumin. For the next two weeks she did not complain, had blood pressures of 150/90 and 140/98, and the urine remained albumin free. She was admitted to hospital at term complaining of headache and vomiting, her blood pressure was 130/90, and a very faint trace of albumin was found in the urine. She was treated conservatively and the blood pressure varied from 140/90 to 170/110 with no albuminuria. One week after admission, with the blood pressure 190/130, labour was induced surgically. The next day her blood pressure was 120/70 and she was noted to be "bright and cheerful." In the afternoon she was allowed to walk around the ward in order to encourage the start of labour. Fifteen minutes later she dropped down dead.

Post-mortem examination showed the pericardial sac to be full of blood. Coarctation of the aorta was present at the origin of the left subclavian artery, and at this point the aorta measured ¾ in. (1 cm.) across. The ascending aorta was dilated to a diameter of 3 in. (7.6 cm.), and showed a split in the intima with a dissecting aneurysm which had ruptured through the adventitia into the pericardial sac. The left ventricular muscle was slightly hypertrophied. A bicuspid aortic valve was present. The uterus contained a normal full-term foetus, and except for congestion of the lungs all other organs were normal.

Case 4

A 1-para 2-gravida aged 24 was walking through the backyard of her home carrying her 15-months-old baby on her arm when she suddenly collapsed, quickly lapsed into unconsciousness, and died after about 30 minutes. No further history was available.

Post-mortem examination showed a small ruptured aneurysm at the bifurcation of the right middle cerebral artery. The subarachnoid haemorrhage was greatest around the optic chiasma, the brain stem, and between the cerebrum and cerebellum. It had tracked along the sulci and was present in the ventricles too. The skull was intact and there were no injuries of the scalp. The heart was slightly enlarged owing to hypertrophy of the left ventricle. The mitral valve showed some fibrosis and scarring, while the aortic valve had two functioning cusps only. Stenosis of the aorta was present at the junction of the arch and descending portions, and measured ½ in. (1.3 cm.) at this point. The uterus contained a normally developed foetus 9½ in. (24.2 cm.) long, corresponding to the pregnancy of 20 to 22 weeks' gestation. Except for venous congestion the remainder of the organs were normal.

Case 5

A woman aged 30 had been attending her local antenatal clinic for her first pregnancy and was said to have enjoyed perfect health both before and during her pregnancy. At about the 38th week she suddenly collapsed at home and died within a few seconds.

On post-mortem examination the pericardial sac was distended with blood which had poured through a small rupture in the posterior wall of the root of the aorta. The ascending and transverse parts of the aorta were very thin and were dilated to a point just behind the left subclavian artery. Here the aorta became narrow and "completely obliterated," while below this point the vessel was noted to be "abnormally narrow." There was marked hypertrophy of the left auricular and left ventricular muscles. The aortic orifice was very narrow and would not admit one finger. The uterus contained a normal 38-weeks foetus, while all other organs were macroscopically normal.

Discussion

The comparatively small number of cases on record do not allow of any valid conclusions of a statistical nature to be drawn from them. Many of the reports are deficient in important details; some are only mentioned in textbooks and articles. Again there is a tendency to report only the unusual and dramatic, and therefore no detailed statistical analysis is possible if an imbalanced picture is to be avoided.

Coarctation of the aorta in an adult carries with it well-defined risks, and the association of pregnancy increases these hazards, especially of rupture of the aorta. There are records of 11 patients with the anomaly whose death was directly associated with pregnancy, and in 6 of them death was due to rupture of the aorta just before term or during labour. In 9 out of the 11 cases the coarctation remained undetected during the pregnancy. Earlier diagnosis, and therefore appropriate supervision and management, may have a large part to play in improving the prognosis. However, perusal of the literature shows that many women have successfully negotiated repeated pregnancies without any demonstrable adverse effects in spite of the presence of coarctation, which in many cases remained undetected until after the pregnancy. One woman of 56 (Weber and Price, 1912) had produced 11 children and died 12 years after her last pregnancy, and another of the same age (Strassmann, 1922) produced seven children. While not diminishing the seriousness with which this association of conditions has to be regarded, these facts suggest that a uniformly gloomy prognosis for all cases is unwarranted. The records, however, give no indication of those women with coarctation of the aorta who are likely to be unaffected by pregnancy—information which might be a useful guide in the management of future patients. Comparative studies of blood-pressure readings or the degree of collateral circulation appear to have no value in deciding prognosis in an individual patient.

Antenatal supervision of pregnancies is now an established practice, and it is usual to examine the chest at least on the first attendance and to measure the blood pressure frequently throughout the pregnancy. These are unusually favourable conditions for detecting those patients who have an existing coarctation of the aorta, and routine palpation of the femoral pulses in all patients with hypertension, however mild, would undoubtedly disclose further examples of the associated conditions.

The advice regarding management of these patients has varied according to the personal experience of the physician and his reading of the literature at the time. Some have advised that all cases should have therapeutic abortion with sterilization (Mendelson, 1940), some have advised caesarean section at term with or without sterilization (Walker, 1943; Baber and Daley, 1947; Brown, 1950), while others have allowed labour to proceed normally (Soulié *et al.*, 1951). Surgical treatment of coarctation of the aorta has not only stimulated early diagnosis of the condition but will now influence the management of pregnancy cases. The patients of Marcorelles (1949) and Peterson *et al.* (1953) were both operated on successfully during their pregnancies, and Miller and Falor (1952) describe the case of a woman who was operated on three months after her first pregnancy and successfully negotiated a second a year later.

It is suggested that all patients in whom these conditions coexist should be referred to a thoracic unit, where the possibility of surgical intervention might be considered. A pregnancy of less than 20 weeks' duration should, in itself, be no contraindication to operation. Until more experience is available it would be wiser to defer surgery if the diagnosis of the coarctation is first made in the later months of pregnancy. Management otherwise is, in general, similar to that applied to pregnant women suffering from rheumatic heart disease. Undue enlargement of the heart, signs of aneurysmal dilatation of the ascending aorta, or symptoms suggestive of cerebral aneurysm indicate the advisability

of terminating the pregnancy. A rise of blood pressure in the last few weeks of pregnancy or during labour should be viewed seriously and caesarean section undertaken.

Summary and Conclusions

There are records of only 91 patients in whom coarctation of the aorta was associated with pregnancy. Five further cases are reported.

The association carries with it serious risks, especially of rupture of the aorta, but, in spite of these, many women have not been adversely affected by multiple pregnancies and have lived to an age well beyond the average expectation of life in coarctation of the aorta. Earlier diagnosis and surgical treatment of the coarctation offer means of improving the prognosis.

Diagnosis is not difficult, and depends mainly on the awareness of the examiner and routine palpation of the femoral pulses in all patients with hypertension. The antenatal clinic offers unusual opportunities for early diagnosis.

General principles in management of these patients are described and the importance of individual assessment and surgical treatment for the coarctation is stressed.

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It is recorded in the annual report of the Chalfont Colony for 1953–4 that an arrangement has been made with the National Hospital for Nervous Diseases whereby patients, either before or immediately after entering the Colony, will go to the Queen Square or Maida Vale Hospitals for full investigation and report. The executive committee believes that a certain percentage of cases now going to the Colony will be intercepted, treated at Queen Square or Maida Vale, and returned to normal life in a few weeks. The medical staff of the Colony will also receive guidance from the staff of the National Hospital about the treatment of those patients who are finally admitted to the Colony.

HAEMORRHAGIC GANGRENE OF THE SKIN IN CHRONIC LYMPHATIC LEUKAEMIA

BY

I. SARKANY, M.R.C.S., L.R.C.P.

AND

RUTH M. RANSOM, M.B., B.Ch., B.Sc.

(From the Peace Memorial Hospital, Watford)

In chronic lymphatic leukaemia both specific and non-specific skin lesions may occur, but distinction between the two often rests on the histological picture.

In a study of the dermatological manifestations of the lymphoblastoma–leukaemia group, Epstein and MacEachern (1937) found that out of 60 patients with chronic lymphatic leukaemia 27 (45%) had non-specific skin lesions. Of these, the haemorrhagic type was commonest (15 patients), and usually occurred in the terminal stages. Although Sutton and Sutton (1941) mention that purpura sometimes results in necrosis and ulceration of the skin and mucous membranes, we are unable to find any reference to the occurrence of haemorrhagic gangrene of the skin in chronic lymphatic leukaemia.

Case History

A 58-year-old male librarian was admitted on October 5, 1952, having been found unconscious in his garden. There was a past history of chronic bronchitis. A provisional diagnosis of subarachnoid haemorrhage was confirmed by lumbar puncture. During routine examination he was found to have moderate splenomegaly but no palpable glandular enlargement, and there were no other abnormal signs. A blood count showed: Hb, 92%; red cells, 4,680,000 per c.mm.; white cells, 12,900 per c.mm. (polymorphs 22%, eosinophils 0.5%, monocytes 1%, lymphocytes 76.5%); no primitive forms; platelets, 168,750 per c.mm.; E.S.R., 1 mm. in the first hour (Wintrobe); clotting-time, 6 minutes 30 seconds; bleeding-time, 3 minutes. A sternal marrow puncture showed a typical picture of chronic lymphatic leukaemia.

As it was uncertain whether the subarachnoid haemorrhage was in any way connected with the leukaemia, and as the leukaemia was symptomless, with the possible exception of being related to the subarachnoid haemorrhage, it was decided not to treat the patient at this stage. He was discharged in November, having made an uneventful recovery from the subarachnoid haemorrhage, and he was seen regularly as an out-patient.

In January, 1953, he was readmitted to hospital with a three-days history of fatigue with fever and the coughing up of sputum flecked with bright blood. On examination he was wasted and there were symmetrical localized areas of ecchymoses over both upper arms and both malar eminences. The only other abnormal physical finding was splenic enlargement, which was of the same degree as on the previous admission. A blood count at this time showed no marked change from the previous count, and the bleeding- and clotting-times were still within normal limits. The possibility of a leukaemoid reaction to generalized tuberculous infection was considered, and accordingly the sternal marrow was re-examined in February, 1953. No evidence of tuberculous histology was seen, and the appearance was still that of a lymphatic leukaemia. A chest *x*-ray examination showed extensive cavitation in the right upper lobe, but repeated sputum tests and examination of laryngeal swabs did not reveal any evidence of tuberculosis.

The skin lesions became more numerous. Each would start as a small patch of purpura which enlarged rapidly