COARCTATION OF THE AORTA AND PREGNANCY

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Pregnancy in a woman suffering from coarctation of the aorta is of especial interest to the obstetrician for several reasons. Its rarity alone would ensure this since only 136 cases have so far been recorded. It is of further interest in that it is the only type of heart disease for which many obstetricians would advocate an elective Caesarean section in preference to a vaginal delivery. Finally, the increasing scope of vascular surgery, with new techniques for excising and grafting the constricted aorta, have brought fresh problems which need to be considered.

The Incidence of Coarctation of the Aorta

The overall estimation for this condition is about 1 in 1,500 people (Blackford, 1928), which makes its rarity in pregnancy at first surprising. Coarctation is, however, four or five times as common in the male (Abbot, 1928; Riefenstein *et al.*, 1947) and in addition, irrespective of pregnancy, these cases may die at an early age, both of which will obviously decrease its frequency in pregnancy. It is probable though that in the past the condition has remained undiagnosed in pregnancy and an increasing awareness would disclose it more frequently. Thus the first case to be found in the Queen Charlotte's Hospital reports was in 1943, then another in 1950 and a third in 1953. At St. Mary's Hospital the first case to be recorded was in 1950, since when there have been another two. The rarity of the condition in pregnancy underlines the importance of reporting these cases when they occur, and this has been emphasized by Novak (1947).

Finally, the further need for detail is emphasized by the difficulty in evaluating any line of management when it is based on such small figures.

Mortality of Coarctation Alone

This is probably in the region of 15 per cent., although reported figures show considerable variation. If death is going to occur, it usually happens before the age of 40. Seventy-four per cent. of Maud Abbott's series (1928) of 200

cases and 61 per cent. of 104 patients reported by Riefenstein *et al.* (1947) died before the fifth decade. The causes of death in these series were: Aortic rupture, 25 per cent.; bacterial endocarditis, 25 per cent.; congestive heart failure, 25 per cent.; cerebral vascular accidents, 10 per cent.; other causes, 15 per cent.

Resection of the aorta has become increasingly common and the operative mortality ranges from 2 per cent. (Gross, 1953) to 10 per cent. (Cleland *et al.*, 1956). It might be noted here that hypertension itself is rarely severe and only exceptionally a cause of death itself. It may, however, be a contributory factor in causing a cerebral vascular accident, especially as congenital 'berry' aneurysms tend to be associated with coarctation.

Hazards of Coarctation in Pregnancy

Goodwin (1958) reported 136 cases, including 13 of his own, of which 10 had been described by Cleland *et al.* in 1956. There are a further 6 cases reported elsewhere not included in this paper. Tebow *et al.* (1954) described 3 cases operated on during pregnancy, and Keswick and Wilson (1954) a further 3, all of which were delivered vaginally with no mortality. To these 6 are added a further 5 of our own, which makes a grand total of 147. In this group there were 13 maternal deaths, giving a maternal mortality of 9 per cent. It would appear that this mortality has been steadily decreasing over the years, but this is probably due to some extent to the fact that many of the early cases were probably reported solely because of the fatality. The mortality related to total number of pregnancies is 13 in 367, which is 3.5 per cent. The cause of death in the fatal cases is as follows: Rupture of the aorta or dissecting aneurysms, 6; cerebral vascular accidents, 2; congestive heart failure and pulmonary oedema, 3; other causes, 2.

It can be seen that nearly half the deaths were caused by aortic rupture and it is worth considering whether pregnancy increases this risk. Rupture of the aorta is usually due to preceding medial necrosis. For example, the fatal case

reported by Kinney *et al.* (1945) showed marked changes in the media proximal to the level of coarctation with much loss of elastic tissue. This author also speculated as to the possible role of hyperlipaemia, especially hypercholesterolaemia, which occurs to a moderate degree in normal pregnancy. Joule (1949) suggested a possible thyroid deficiency as being important due to a failure of the gland to meet the excess requirements demanded during pregnancy. It would be imagined that this weakness would become more evident when associated with hypertension, but this has not been a marked feature of those cases that did actually rupture (Riefenstein *et al.*, 1947).

Furthermore, the evidence from Dixon and Hartley's cases (1955) is that the blood pressure behaves very much as in normal pregnancy, dropping in the middle trimester before returning to its former level. The records of 3 of our cases showed this quite clearly. It might then be thought that the lability of the blood pressure could be the precipitating cause, but although this is a feature of all patients with coarctation it is not particularly evident in pregnancy. Whereas the effects of pregnancy on a patient suffering from coarctation alone are a matter for debate, if there is superadded heart disease then the prognosis is definitely worsened. The increased cardiac output that occurs in pregnancy will accentuate any failure present, due say to an associated aortic incompetence. This complication is, however, fortunately rare.

One of the cases delivered at Queen Charlotte's Hospital had a subsequent resection at Hammer-smith Hospital and is included in Goodwin's series and will not therefore be described here.

Case Reports

Case 1. Mrs. J.R., aged 33, attended Queen Charlotte's Hospital in 1943 for her third pregnancy. The first, in 1929, had been delivered by forceps and the child weighed 7½ lb. In 1935 the patient had had a full-term breech delivery but this child was stillborn and weighed only 4 lb.

Past medical history was uneventful and she had not complained of any dyspnoea or other symptoms at her first attendance. Her blood pressure on this occasion was 185/100 and examination of her cardiovascular system showed definite cardiac enlargement with a strong impulse and systolic murmurs in both mitral and aortic areas. The femoral pulses were absent. X-ray of her chest showed Roesler's sign on both sides and screening confirmed the diagnosis of coarctation.

Her ante-natal attendances were without incident, although her hypertension persisted to a varying degree throughout the pregnancy. An

external cephalic version was performed without an anaesthetic at the 38th week. Delivered at term normally of a 6 lb. 8 oz. child, first stage lasting 13 hours and the second 10 minutes. The puerperium was quite normal.

She was referred to a cardiologist for further follow-up, but failed to keep her appointment.

Case 2. Mrs. B.R., aged 26. This patient's past medical history was also uneventful. She had had one child when aged 20 and this had been delivered with difficulty by forceps at term and weighed 7 lb. 12 oz. It was noted during this pregnancy that she had a mild hypertension. Her booking blood pressure was 135/80 and rose terminally to 150/100. It was thought then by the cardiologist who saw her that she had some form of congenital heart disease, but no further details were forthcoming.

When she attended Queen Charlotte's Hospital for this pregnancy her blood pressure was noted to be 170/80 and examination of her cardiovascular system showed a slight enlargement of the left ventricle with a loud systolic murmur all over the praecordium. Anastomotic vessels were present on her back and her femoral pulses were absent.

Her ante-natal progress was normal, but pelvimetry confirmed a degree of outlet contract and she was delivered by an elective lower segment Caesarean section at the 39th week of a healthy 7-lb. child. Her recovery was uninterrupted and she was discharged on the 16th post-operative day. Her blood pressure at the follow-up clinic was noted to be 140/110.

Case 3. Mrs. V.W., aged 37. The diagnosis of coarctation of the aorta had been made five years prior to the present pregnancy on the findings of weak pulses in the left arm and both femoral arteries, absent pulsation in popliteals and arteries of foot, rib notching, a widely conducted systolic murmur and evidence of left ventricular hypertrophy. At the time of the original diagnosis she had an infective endarteritis, which responded to Penicillin.

During the ante-natal period of the present pregnancy the blood pressure was raised in the right arm at nearly all visits, the maximum being 170/110. The blood pressure in the left arm was on all occasions below 140/90. The patient was delivered at the 39th week by elective lower segment Caesarean section of a living male child weighing 7 lb. The puerperium was uneventful. The only indication for abdominal delivery in this case was the presence of coarctation of the aorta.

Case 4. Mrs. G.S., aged 25. She was known to have a 'murmur' as a child. Coarctation of the aorta was diagnosed at the age of 19 on the findings of hypertension (240/95 in the right arm,

FIG. 1.—The terylene cloth prosthesis in position.

245/100 in the left arm), absent femoral pulses, apical systolic murmur also audible between scapulae, periscapular pulsation. Further investigation revealed an aneurysmal dilatation just distal to the constriction. Her first pregnancy in 1950 was terminated because of the coarctation and associated aneurysm. She became pregnant again in 1955 and first attended the ante-natal clinic. Because of the aneurysmal dilatation of the aorta distal to the coarctation, and in view of the degree of hypertension, it was decided to operate despite the pregnancy. The coarctation and aneurysm were excised and continuity in the aorta re-established by a tubular terylene cloth prosthesis.

The patient had a complete miscarriage 48 hours after operation. She made a good recovery from the operation. Femoral pulses became palpable but the hypertension persisted.

During the present pregnancy her blood pressure varied between 150/70 and 180/100. She was admitted for rest and sedation at the 36th

week because albuminuria and oedema had appeared. The blood pressure on admission was 190/100. The hypertension continued during the next three weeks, but the albuminuria was present only occasionally. Delivery was by elective lower segment Caesarean section at the 39th week. It was noted at this time that her abdominal aorta was well developed although slightly smaller than normal and pulsating freely. The child was alive and weighed 9 lb. 1 oz. The obviously large baby and the presence of an inert graft in the aorta were the chief factors determining the abdominal delivery.

Case 5. Mrs. J.P., aged 22. At the age of 15 the patient attended the medical out-patient department complaining of frontal headaches, blurring of vision and coldness of her lower extremities. A diagnosis of coarctation of the aorta was made in view of the findings of hypertension (180/100 in the right arm, 160/100 in the left arm), absent femoral pulses, apical systolic murmur, periscapular arterial pulsation and rib

notching on chest x-ray. In addition, the patient had moderately well-developed webbing of the neck. She was treated by excision of the affected segment of the aorta below the origin of the left subclavian artery together with ligation of a patent ductus arteriosus. Continuity in the aorta was obtained by end-to-end suture. She made a good recovery following the operation and the improvement in her symptoms was maintained. She became normotensive and good arterial pulses were present in both lower limbs.

During the present pregnancy the ante-natal period was uneventful until the 30th week when hydramnios occurred. The blood pressure at the 9th week was 110/75, and it remained normal until the 32nd week when it was found to be 164/106 and was associated then with gross generalized oedema and albuminuria. She was treated by bed rest, heavy sedation and salt restriction. During the next four days her condition deteriorated. There was oliguria, increasing albuminuria to 4 g. per litre, and hypertension rising to a maximum of 170/125. In view of this the pregnancy was terminated at the 32nd week by lower segment Caesarean section. The child weighed 3 lb. 7 oz., was born alive but died after 30 minutes. Delivery was followed by a massive diuresis. The urine became protein-free by the 6th week post-partum. The hypertension did not settle spontaneously and she was eventually rendered normotensive by Serpasil 0.75 mg. and Inversine 25 mg. daily in divided doses.

Discussion

It is evident from the weight of reported evidence that pregnancy in patients with coarctation of the aorta can be allowed to continue to term without any increase in the mortality rate which is due to the coarctation alone. The view expressed by Mendelson (1940) that pregnancy should be terminated if diagnosed early enough has not found support in recent years, except in those occasional cases where there is a serious associated cardiac lesion.

Interest has been aroused recently over the problem of the safest route of delivery for these patients. Benham (1949) reviewed the previously recorded cases, added three of his own, and concluded that delivery should be by Caesarean section in order to avoid the rise of blood pressure in labour, which is agreed to occur by nearly all observers. He further quotes the observation of Cook and Briggs (1903) that a similar rise may occur during instrumental delivery or vaginal manipulation under anaesthesia. Pritchard (1953) considered that delivery should be by the vaginal route as he had found a comparable mortality rate when comparing abdominal and vaginal routes of

delivery. It must, however, be noted that these mortality rates were calculated on a very small number of cases with only two deaths in the vaginal, and one in the abdominal series.

Dixon and Hartley (1955), as a result of their observations on two cases, found no evidence of a rise in blood pressure during delivery, favoured vaginal delivery. Goodwin (1957), reviewing the previously reported cases and adding 13 of his own, favoured vaginal delivery with a shortened second stage, although the only case which he reports in detail was delivered by Caesarean section in two pregnancies.

Operative treatment of coarctation is likely to remove patients from the risk of aortic rupture or dissection, and of congestive cardiac failure. Because of the presence of a healed anastomotic scar or inert graft in the aorta, they are, however, likely to continue to be at risk from endo-aortitis, or endo-carditis where there is an associated valvular defect. Also, the presence of congenital aneurysms of cerebral arteries in association with coarctation means that surgical treatment of the latter will not necessarily prevent death from cerebro-vascular accidents, although reduction of blood pressure in adolescence will reduce the liability of development, or subsequent rupture of these aneurysms (Campbell and Baylis, 1956).

Although the evidence is not entirely clear, it must be a logical assumption that a rise of blood pressure would be dangerous for patients with an unresected coarctation and to a lesser extent when resection has been performed. We consider that it has not been clearly established that a labour with the second stage shortened by the use of forceps can prevent this rise. One of the cases described by Tebow (1954) which had been treated by resection showed this rise quite clearly. In addition, the use of forceps to shorten the second stage may be a simple procedure, but in primigravidae this is not always so and may entail more manipulation and difficulty than first expected. Similarly, the first stage itself may be protracted and it is not possible to guarantee an 'easy' delivery without being ready to perform a Caesarean section rather sooner than is normally indicated.

If this should have to be done, it would appear that these patients have got the worst of both worlds. A further objection to the use of forceps to shorten the second stage of labour in these patients is that strict asepsis cannot be obtained in vaginal operations and that such a method of delivery would, in our view, add to the risk of endo-aortitis or endocarditis, about which all writers on this subject are most concerned. We consider that Caesarean section, carried out electively, and with the full aseptic technique avail-

able in an operating theatre, is the optimum route of delivery for these patients, thereby avoiding altogether the possible rise of blood pressure in labour and delivery and lessening the risk of subsequent endo-aortitis. This is the view shared by Benham (1944), Bramwell (1953), and Rosenthal (1955). The latter writer comments that 6 out of the 11 cases of pregnancy and coarctation which terminated fatally died in labour or near term.

While advocating an elective Caesarean section for primigravidae, where the condition is met after a previous normal vaginal delivery, we suggest that a further vaginal delivery could be allowed when a short first stage of labour and a rapid normal second stage can confidently be expected. The first two case reports show this carried out in practice. The first had had two deliveries, there was no evidence to suggest any expected obstetric difficulty, and the second stage in fact lasted 10 minutes. The second patient gave a history of a difficult forceps delivery, there was some outlet contraction present, so a Caesarean section was decided on.

In addition to this proposed method of delivery, it is obvious that patients with coarctation must receive careful ante-natal care, periods of rest if cardiac symptoms arise, and antibiotic prophylaxis in the puerperium as advocated by MacLeod (1954).

Two of our cases developed pre-eclamptic toxæmia in the last trimester of their pregnancies. It is surprising that this added hazard has not been noted more frequently in view of the higher incidence of toxæmia that occurs in patients suffering from essential hypertension. If, as some authorities suggest, the hypertension of coarctation is a product of renal ischaemia, then these patients would be expected to be very liable to develop pre-eclamptic toxæmia. It is also of interest to note that in both these cases the coarctation had been resected.

Although resection during pregnancy has been described several times, Pritchard (1953) emphasized the risk to the foetus from prolonged anoxia and this is borne out by the history of Case 4,

where abortion occurred 48 hours after operation.

Webbing of the neck is a characteristic feature of the syndrome of gonadal dysplasia described by Turner (1938) and Albright *et al.* (1942), and coarctation is sometimes found as an additional feature. Grumbach (1955) described 22 cases of gonadal dysplasia with 12 showing webbing and coarctation and found them all to be chromosomal males, which is another typical feature of the syndrome. Case 5 where webbing and coarctation were present and obviously not associated with gonadal dysplasia must therefore be of exceptional rarity.

Summary

The incidence and hazards of coarctation of the aorta associated with pregnancy are discussed. Five case reports are presented and the obstetric management described. The conclusion is drawn that, in general, these patients are best delivered by an elective Caesarean section.

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