Coarctation of the aorta corrected during the first month of life

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SUMMARY Forty eight children (29 boys) had surgical correction of coarctation of the aorta during the first month of life; all had patent ductus arteriosus. The 33 survivors were reviewed at a mean age of 6.6 years. Of the 19 children with no associated anomaly, none had died. The more complex the associated anomalies, the greater the mortality. Two (6%) of the survivors, both with associated anomalies, have some residual disability; one is incapable of leading an independent life. No survivor has systemic hypertension. Six (18%) of the survivors have required correction of recurrent coarctation, and one is awaiting repair.

Newborn babies suspected of having coarctation should be assessed for surgical correction without delay, and medical treatment (including, if necessary, infusion of prostaglandin E₂ in a dose of 0.025 µg/kg/minute) should be instituted in the interim. Long term follow up is important to detect systemic hypertension or recurrence of the coarctation. This occurred in seven (21%) of our survivors.

Coarctation of the aorta presenting in the neonatal period, with or without associated anomalies, is a life threatening condition, and the prognosis is poor if surgical correction is not carried out. Recent advances in medical management, in particular infusion of prostaglandin E₂, permit such babies to be stabilised in better condition before operation. Despite such advances, however, many babies arrive in the cardiothoracic surgical unit with severe complications—for example, cardiac failure, hypoxia, acidaemia, shock, renal failure, or septicaemia.

The immediate results of surgical correction have been reported from a number of centres including our own.^{1–4} Potential long term complications include recurrence of the coarctation, the effects of any coexisting anomalies, the consequences of any perioperative problems, and the development of systemic hypertension.

We present our results in 48 consecutive babies with coarctation of the thoracic aorta who underwent surgical correction during the first month of life and were reviewed in childhood.

Methods

Correction of coarctation of the aorta was carried out in 48 babies aged 1 month or less between March

1978 and April 1982. Subclavian flap correction⁵ was carried out in 45 children, each of the remaining three having a polytetrafluoroethylene patch prosthesis inserted. All 48 children had patent ductus arteriosus.

For the purposes of this study we allotted the children to one of three groups according to the severity of their coexisting anomalies. The first group had no associated intracardiac lesion, the second group had associated ventricular septal defects, and the third group had more complicated anomalies.

At follow up the children were assessed according to recurrence of the coarctation, the development of systemic hypertension, their general physical health, and any evidence of handicap.

Results

The clinical features of the 48 babies are shown in the table. There were 29 boys and 19 girls, and all had patent ductus arteriosus.

BABIES WITH NO INTRACARDIAC ANOMALY

Twelve boys and seven girls had no other anomaly; their gestational ages ranged from 36 weeks to 41 weeks 3 days, and their birth weights from 2290 g to 3540 g. There were no deaths in this group, and they

Table Features of 48 babies with coarctation of the aorta operated on during the first month of life.

	Associated anomalies		
	None	Ventricular septal defect	More complex anomalies
Total No	19	15	14
Sex (male:female)	12:7	8:7	9:5
Mean (SD) birth weight (g)	3050 (360)	2770 (680)	3380 (680)
Mean (SD) gestational age (weeks)	39.2 (1.8)	38.1 (2.2)	40.6 (1.2)
Mean (SD) age at operation (days)	14.2 (7.6)	10.3 (6.5)	12.0 (6.7)
No of early deaths	0	5	5
No of late deaths	0	0	5
No of survivors	19	10	4
Sex (male:female)	12:7	5:5	2:2
Mean (SD) age at follow up (years)	6.70 (1.47)	5.96 (1.15)	7.45 (0.96)
No of recurrences of coarctation	8*	1	1†
Mean (SD) systolic blood pressure (mm Hg)	117 (15)	101 (17)	103 (9)
Mean (SD) diastolic blood pressure (mm Hg)	70 (8)	60 (6)	64 (10)
Disability	0	1	1

*Coarctation recurred twice in one boy and one girl; †this boy, with a single ventricle, had second repair when 11 months old and died at the age of 13 months.

were reviewed at a mean age of 6.7 years (range 4.6 to 9.3).

Four children required a second correction of coarctation at the ages of 1.3 years, 2.4 years, 4.4 years, and 6.4 years, respectively. The first and third of these children each required a third correction at the ages of 8.1 years and 6.5 years, respectively, and one further child awaits correction of a recurrence at the age of 6.2 years.

Apart from the child who is waiting for correction of a recurrence no child has a disability in this group. All the children are in good physical health without systemic hypertension, and are developing normally.

BABIES WITH AN ASSOCIATED VENTRICULAR SEPTAL DEFECT

Eight boys and seven girls had only an associated ventricular septal defect. Their gestational ages ranged from 33 to 40 weeks, and birth weights from 1770 g to 4010 g. Five babies (33%) died between one and 28 days postoperatively, three in 1978 before the introduction of prostaglandin E₂ infusion to improve their preoperative condition.

One child (7% of the original group), a girl of 6.4 years, is disabled. She weighed 2800 g at term, and

was acidaemic and severely ill when she presented. Postoperatively she had several seizures. She had a renal vein thrombosis leading to chronic renal failure, and has failed to thrive. She is now being considered for a renal transplant, and attends a school for children with learning difficulties.

The remaining nine children (60% of the original group) are well at a mean age of 5.9 years (range 3.8 to 7.4) and are developing normally. One girl required correction of a recurrent coarctation at the age of 3 months. None of the 10 survivors has systemic hypertension or further recurrence of the coarctation, and none has required an operation for the ventricular septal defect.

BABIES WITH MORE COMPLEX ASSOCIATED ANOMALIES

Nine boys and five girls had more complex associated anomalies. Their gestational ages ranged from 38 to 43 weeks, and their birth weights from 2400 g to 4800 g. Five babies (36%) died between five and 28 days postoperatively. All were expected to require eventual intracardiac repair. Five (36%) died between the ages of 3 months and 3.2 years. In each case, intracardiac repair had either been undertaken or was planned. There were four survivors (29%).

One boy aged 7.5 years with aortic stenosis weighed 3790 g when he was born at full term. He required treatment with digoxin and frusemide before operation, which was carried out when he was 8 days old, and after operation he was treated with phenobarbitone because of some twitching. He now attends a school for children with moderate learning difficulties. His blood pressure is 105/50 mm Hg and he has had no recurrence of the coarctation.

A girl of 5.9 years with aortic stenosis weighed 2380 g when she was born at 38 weeks' gestation. After operation at the age of 6 days she has progressed satisfactorily, growing with height and weight at the third centile, blood pressure 100/75 mm Hg and no recurrence of the coarctation.

A boy of 7.9 years with a ventricular septal defect, mitral stenosis, and aortic stenosis weighed 3010 g when born at full term. After operation at the age of 22 days he has grown at the 25th centile for height and weight, has had no recurrence of the coarctation, his blood pressure is 115/70 mm Hg, and on echocardiography there is normal left ventricular function, normal pulmonary artery pressure, trivial aortic incompetence, and an immobile posterior mitral leaflet.

A girl of 8.5 years with a ventricular septal defect, hypoplastic left ventricle, and hypoplastic mitral valve, weighed 4260 g when born at full term. She was in severe cardiac failure at presentation, was treated with digoxin and frusemide, and had several

seizures after the operation, which was carried out when she was 9 days old. Her height is at the 90th centile, her weight at the 97th centile, her blood pressure is 90/60 mm Hg, and she has had no recurrence of the coarctation.

Discussion

Coarctation of the aorta is a common congenital cardiovascular malformation, and occurs more often among boys. In this series (from a supraregional cardiothoracic surgical centre) there was a male:female ratio of 1.5:1.

Newborn babies with critical aortic coarctation become symptomatic when the patent ductus arteriosus closes. Before this the femoral pulses may be easily palpable because of ductal blood flow. Ductal closure occurs at a variable time after birth, and our babies presented at ages starting at 1 day but the peak incidence occurred at about the age of 1 week. All the babies presented with cardiac failure.

The diagnosis of coarctation of the aorta should be considered in any baby in cardiac failure. Physical signs may include a systolic murmur, but the important finding is a difference in blood pressure (using an appropriately sized cuff) between the right upper limb and the lower limbs. This important sign will not be present in the rare case where there is an aberrant arterial supply to the right upper limb, if the supply originates beyond the coarctation when the right upper limb pressure is also low.

Babies in whom the diagnosis of coarctation of the aorta is suspected should be referred without delay to a cardiothoracic surgical centre with paediatric experience where definitive diagnosis can be made, generally by echocardiography, before urgent surgical correction.⁶ Appropriate medical management should be instituted promptly while the transfer is being effected. After discussion with the staff at the referral unit, if the baby's condition is deteriorating or if there is likely to be substantial delay before transfer, an infusion of prostaglandin E₂ should be started. Our starting dose is 0.025 µg/kg/minute given by intravenous infusion. Larger doses are more likely to lead to episodes of apnoea or bradycardia. If the baby is in a poor haemodynamic state the merits and risks of adding dopamine to the infusion should be discussed with the staff at the referral unit.

The prognosis is determined primarily by the severity of any coexisting anomalies.² In our series there was no mortality if there were no associated

anomalies. Of 15 babies with ventricular septal defects five died, all in the immediate postoperative period; three of these deaths, however, occurred in 1978 before we had started to use prostaglandin E₂ infusions. On the other hand, only four children have survived of the 14 who had more complicated associated anomalies.

Any complications, whether before, during, or after operation, influence mortality and morbidity. The introduction of intravenous prostaglandin infusions permitted more babies to have their operations without being in cardiac failure, or hypoxic, or acidaemic, or shocked, and is likely to have favourably influenced outcome.

There is debate about the best surgical technique,^{7 8 10} and our experience may contribute to that debate. Whatever method of repair was used, we recommend continuing follow up of all patients, who may have recurrence of the coarctation (as occurred in 21% of our survivors) and who may develop systemic hypertension.⁹

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