Coarctation of the Aorta in Infants Under One Year of Age

WITH PARTICULAR REFERENCE TO RESULTS OF SURGERY

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SUMMARY

During the 10-year period 1962 - 1971, coarctation of the aorta was diagnosed within the first 5 months of life in 35 hospital cases. Of these, 29 (83%) were symptomatic, and 18 (54%) underwent surgery to correct the coarctation. Thirteen of the 18 patients (72%) survived the procedure. Of the 5 patients who died, 2 had single-ventricle complexes, and 1 had an associated ventricular septal defect and died at a subsequent operation for pulmonary artery banding. One patient who survived had a thoracotomy with no procedure done to the aorta.

All survivors were followed up for at least 1 year. Residual gradients were found in 6 of the 12 patients (50%), but classified as severe in only 2 cases.

Of the 11 patients who were symptomatic but who did not undergo surgery, 7 died (mortality 63%). There were 6 remaining patients who were asymptomatic. There have been 3 deaths in this series—all unrelated to their cardiac pathology.

It is strongly recommended that young babies with coarctation of the aorta, who develop congestive cardiac failure, undergo 36 - 48 hours of medical therapy, after which surgical resection of the coarcted segment is carried out. This approach offers the best prospects for survival.

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There is very little controversy as to the management of coarctation of the aorta in infants who present early with heart failure.[1,2] Authors agree that surgery plays an important role after the institution of medical therapy, and is indicated in any infant with coarctation of the aorta who has not responded completely within 36 - 48 hours to medical therapy.

We report a study of infants under the age of 1 year with coarctation of the aorta, with particular reference to the results of surgical management.

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PATIENTS

All infants with coarctation of the aorta who were under 1 year of age when admitted to the Transvaal Memorial Hospital for Children during the 10-year period, January 1962 to December 1971, were studied. There were 35 cases. Babies with associated cardiac and non-cardiac anomalies were included in the study.

The cases that were symptomatic and in congestive cardiac failure (CCF) were at first treated medically for 36 - 48 hours with digoxin. In addition, oxygen, the maintenance of fluid and electrolyte balance, and usually a diuretic (intramuscular furosemide 1 mg/kg) were necessary. The infants were then submitted for resection of the coarctation. The reason for this approach was our previous experience with a patient who relapsed and died after initial improvement on medical therapy alone.

The surviving patients have been followed up for 1 - 7 years and the question of residual coarctation, or 're-coarctation' has been reviewed. Careful palpation of the femoral pulses was carried out and flush blood pressures were recorded in the upper and lower limbs.

RESULTS

The age at diagnosis is shown in Fig. 1. Twenty-four of the 35 cases (68%) were seen in the first 3 weeks of life, the remainder before the 5th month of life. Fig. 2 shows the ages of the patients who were operated upon. From Fig. 3 it can be seen that the patients have been subdivided into 3 groups. In group I thoracotomy was performed on 18 cases, because of heart failure. There was a 72% survival rate (13/18) from the operation, although in 1 case nothing definitive was done to the infant because of an associated tubular hypoplasia of the aorta (but the infant survived).

There were 17 patients who were not operated upon, ot whom 11 were symptomatic (group II). Seven of the latter group (64%) died before they could be operated upon. Of the remaining 4 patients who survived, 3 were in mild congestive cardiac failure and improved rapidly within 24 hours of digitalisation. The 4th patient presented with cyanosis arising from a common atrioventricular canal, and in whom the coarctation had been missed.

There were 6 asymptomatic infants whose coarctations were detected incidentally (group III). Of these, 3 died (50%), but the causes of their deaths were not cardiac (1 patient died from *E. coli* meningitis, 1 from complica-

tions of hyperalimentation after gastro-enteritis, and the third was an infant weighing 1 kg, who died of *E. coli* septicaemia).

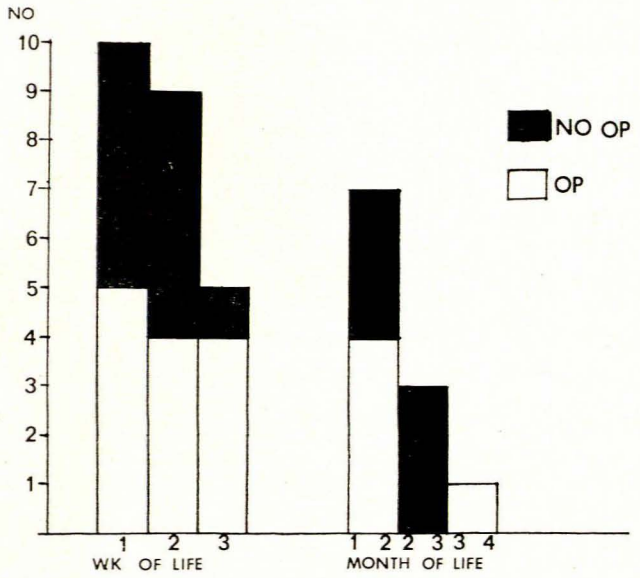

Fig. 1. Age of infants at diagnosis of coarctation of aorta (operated and not operated).

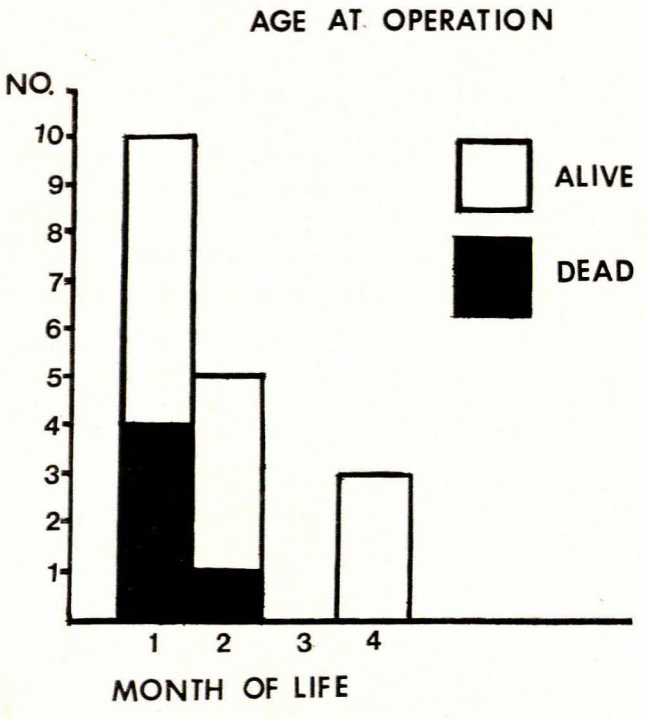

Fig. 2. Ages of 18 infants who underwent surgery for coarctation of aorta.

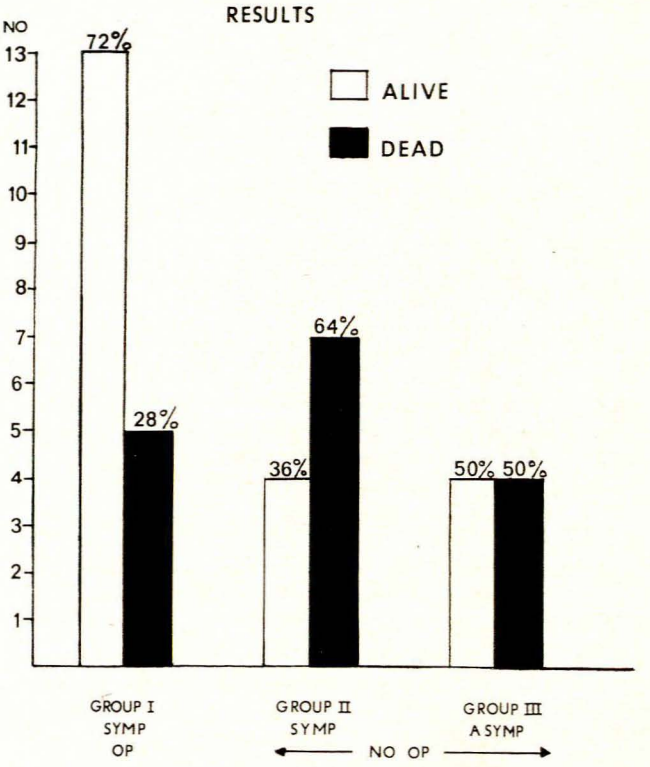

Fig. 3. Over-all results in 35 infants with coarctation of aorta. Symp.—symptomatic; Asymp.—asymptomatic; Op.—operation; No op.—no operation.

Analysis of Deaths in Groups I and II

Of the 5 fatal cases in the group operated on (all deaths were early), 2 had single-ventricle complexes, 1 of these being associated with a corrected transposition of the great vessels. The other deaths occurred as follows: in 1 patient the coarctation was not completely relieved; 1 patient died from poor postoperative management; and the last patient died after a third operation to band the pulmonary artery for pulmonary hypertension caused by an associated ventricular septal defect (VSD). This baby survived the first thoracotomy for resection of the coarctation and a repeat coarctectomy for residual gradient.

TABLE I. COARCTATION IN FIRST TWO WEEKS OF LIFE (SYMPTOMATIC)

	Alive	Dead	Total
Operated on	**6**	**4**	**10**
Not operated on	**0**	**5**	**5**
Total	**6**	**9**	**15**

The 7 deaths in group II were associated with VSD and patent ductus arteriosus (PDA) in 3 cases; PDA alone in 2 cases; a possible fibro-elastosis in 1 case (no post-mortem examination); and a bicuspid aortic valve in 1 case.

TABLE II. ASSOCIATED ANOMALIES IN 35 CASES

	No operation	Operation	Total
Cardiac			
PDA	6	13	19
VSD	6	4	10
Hypoplasia of arch	1	3	4
Single ventricle	—	2	2
Aortic valve anomaly			
(a) AS	1	1	2
(b) Bicuspid	—	1	1
Anomalous rt subclavian artery	1	1	2
Corrected transposition	—	1	1
ASD	1	—	1
AV communis	1	—	1
Fibro-elastosis	1	1	2
Pulmonary stenosis	—	1	1
None	2	—	2
Total	20	28	48
Non-cardiac			
Turner's syndrome (XO)	2	—	2
Edward's syndrome			
(trisomy E)	1	—	1
(trisomy 16 - 18)	1	—	—
Total	3	—	3

AS—aortic stenosis; ASD—atrial septal defect; AV—atrioventricular.

Early Cases

On reviewing the outcome of 15 cases who presented with symptoms within the first 2 weeks of life (Table I), 6/10 survived after operation, but none survived without surgery.

TABLE III. RESIDUAL GRADIENTS IN 12 CASES OPERATED ON

	Systolic BP	No.
No gradient	(0 - 20 mmHg)	6
Mild-moderate gradient	(21 - 40 mmHg)	4
Severe gradient	>40 mmHg	2
Total		12

ASSOCIATED ANOMALIES

Associated cardiac abnormalities were extremely common in all 3 groups, and are listed in Table II. VSD and PDA, singly or in combination, were most frequent (29 instances). There were 3 chromosomal anomalies (2 XO and 1 trisomy 16 - 18).

Residual Gradients

From Table III the residual gradients of 12 out of 13 cases operated on, can be seen. The patient with associated tubular hypoplasia, who had nothing done to the aorta at operation, is excluded.

Six (50%) had gradients of 20 mmHg, or more. Only 2 of these had severe gradients (over 40 mmHg). One of these had mild hypertension in the upper limbs. No other patient had hypertension. All gradients were noted very shortly after operation, except in 1 patient in whom a mild gradient developed. None of the infants subsequently developed CCF.

DISCUSSION

From this series it would appear that surgery is a very useful form of therapy for infants with coarctation of the aorta, who develop heart failure. Our results compare favourably with most reported series,[1,3,4,6,8,9] but not all[5,7,10,11] (Table IV). It is the case complicated by aortic

TABLE IV. MORTALITY OF MEDICAL *v.* SURGICAL THERAPY FOR COARCTATION (UNDER 1 YEAR OF AGE)

		Medical		Surgical	
Author	Year	No.	%	No.	%
Sinha *et al.*[2]	1969	18/26	69	10/28	36
Litwin *et al.*[1]*	1971	—	—	14/46	30
Mortensen *et al.*[3]	1959	4/8	50	0/8	0
Glass *et al.*[4]	1960	37/74	50	14/34	41
Freundlich *et al.*[5]	1961	12/27	44	3/3	100
Malm *et al.*[6]	1963	—	—	6/12	50
Lindesmith *et al.*[7]	1966	—	—	3/5	60
Hartman *et al.*[8]	1967	—	—	5/15	33
Tawes *et al.*[9]	1969	—	—	81/179	45
Adam *et al.*[10]	1967	—	—	30/59	51
Chang and Burrington[11]	1972	31/53	58	30/57	53
Present series		7/11	64	5/18	28

* Infants under 2 years of age.

TABLE V. RESIDUAL GRADIENTS IN INFANTS UNDER 1 YEAR OF AGE OPERATED ON FOR COARCTATION

Author	Gradient					
	Mild-moderate		Severe		Total	
	No.	%	No.	%	No.	%
Litwin *et al.*[1]*	**6/30**	**20**	**4/30**	**14**	**10/30**	**33**
Sinha *et al.*[2]					**7/18**	**39**
Tawes *et al.*[9]					**19/98**	**19,5**
Ibarra-Perez *et al.*[12]	**5/12**	**42**	**1/12**	**8**	**6/12**	**50**
Pelletier *et al.*[13]			**5/13**	**38**	**5/13**	**38**
McNamara and Rosenberg[14]	**16/36**	**44**	**14/36**	**39**	**30/36**	**86**
Hartman *et al.*[15]	**8/20**	**40**	**4/20**	**20**	**12/20**	**60**
Eshaghpour and Olley[16]	**8/20**	**40**	**4/20**	**20**	**12/20**	**60**
Present series	**4/12**	**33**	**2/12**	**17**	**6/12**	**50**

Infants under 2 years of age.

hypoplasia and associated VSD, that does not do well. It may be argued that it is the more complex case that dies unless operated on, but from our experience, there is no difference between the cases operated on and not operated on as regards associated cardiac anomalies, with the possible exception of single-ventricle complex.

The ability to resuscitate the sick infants before operation was the determining factor for survival in most instances. The pre-operative management of the infant who is, possibly, hypothermic or acidotic, and who may have been transported to hospital under unfavourable circumstances, is of the utmost importance. Even with intensive care, these infants may not survive.

From other studies (Table V) postoperative residual coarctation of significant degree was present in 18 - 38% of cases, and up to 86% may have some form of gradient subsequently.[1,2,9,12-16] This occurred in half the survivors in our small series. The mechanism of production of the gradient has been thought to be either thrombus at the suture line (gradient noted postoperatively), or fibrosis at the line of sutures (gradual or rapid development of gradient). From other studies it appears that the method of anastomosis is not important in the development of gradients. In addition, it would seem that 5 out of 6 of our cases had immediate postoperative gradients, but they were probably not always due to thrombosis, but possibly due to incomplete surgical relief of the coarctation.

A residual gradient appears to be acceptable as a long-term complication after life-saving surgery.

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