Management of Coarctation of the Aorta in Infants and Children: Preferential Use of Subclavian Flap Aortoplasty

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SUMMARY

From July 1979 to July 1983, 40 patients with coarctation of the aorta were operated upon. Twenty-five were less than 1 year of age, with 16 less than 1 week old at surgery. In 30 patients, the subclavian flap technique was utilized for repair, while various techniques were utilized in the other 10 patients. Seventeen patients had isolated coarctation of the aorta, including all 14 patients older than 1 year at surgery. Associated intracardiac anomalies were present in the other 23 patients, including all patients less than 6 months old. Five of the 16 patients less than 1 week old died; all had associated severe intracardiac anomalies, 4 with some varient of hypoplastic left heart syndrome. There was only one death late among the 24 patients older than 1 week, in an 11 month old with grade IV pulmonary vascular disease.

This data indicates that repair of coarctation of the aorta can be safely performed at any age or size; the mortality is related to any associated intracardiac anomalies. Thirty-four survivors continue to do well up to 4 year postoperatively. None has an arm-leg gradient greater than 10 mmHg and repeat catheterization in 9 patients demonstrates satisfactory growth. Thus the subclavian flap repair remains our technique of choice for coarctation of the aorta, especially in tiny neonates.

INTRODUCTION

Patients with coarctation of the aorta present with absent or diminished lower extremity pulses, either early in infancy with severe congestive failure or later in life, usually with asymptomatic hypertension. Those patients presenting with failure in infancy have a significant associated cardiac lesion in 67% to 95% cases, most frequently a PDA, VSD, bicuspid aortic valve or mitral valvular disease. Medical management of the congestive failure alone is associated with a 40 to 100% mortality (Glass, Mustard and Keith 1960, Kilman et al 1972), prompting an aggressive surgical approach in this group of patients.

Resection of the coarctation with end-to-end anastomosis remains the traditional method of repair. However, reports of a high incidence of recoarctation, especially in patientsoperated in infancy, have led to the development of other surgical techniques. In 1966, Waldhausen and Nahrwold introduced the "subclavian flap" method, in which the coarctation membrane is excised and the transected subclavian artery is turned down as a patch across the narrowed aortic segment. We are now utilizing this method in the repair of coarctation of the aorta in almost all infants and children unless existing anatomy precludes its use.

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CLINICAL MATERIALS AND METHODS

In a 4 year period, 40 patients underwent repair of coarctation of the aorta. Five patients ranging in age from 18 months to 19 years presented recurrent coarctation following initial repair elsewhere. Nine patients had ages ranging from 2 to 16 years. Of the remaining 26 patients, 16 were less than 1 week old, and 14 were less than 3 days of age (Table - I).

TABLE – I
REPAIR OF CORCTATION OF THE AORTA
UNIVERSITY OF MARYLAND HOSPITAL
July 1979 – July 1983

	No.	Weight	Male	Female
Total Series	40	14–42.0 (6.4)	24	16
W 1 Year	24	14–6.0 (3.7)	15	11
W 1 Month	20	1.4–4.0 (3.2)	10	10
W 1 Week	16	1.4–4.0 (3.1)	7	9
W 3 Days	11	1.9–4.0 (3.0)	6	5

In all cases diagnosis was confirmed by cardiac catheterization/angio-cardiography/aortography. In the total series, 17 patients had coarctation of aorta as an isolated lesion. Twenty-three patients presented with coarctation in association with one or more congenital cardiovascular defects: PDA (9), VSD (11), TGA (3), aortic stenosis (4), mitral atresia (3), tricuspid atresia (1), AV canal (1), ASD (1), aberrant right subclavian artery (1). (Table - II).

When presenting with an indication for repair, all patients with coarctation were operated upon, regardless of the complexity of the associated defects or the gravity of the patient's clinical status.

TABLE
TABLE – II
MAJOR ASSOCIATED LESIONS
Among 25 Infants

None	5 (23%) *
PDA Alone	2
ASD	1
Single VSD	4
Multiple VSD's	3 **
Aortic Stenosis	2 **
Hypoplastic Left Ventricle	2
TGA (Taussing–Bing 1)	4 +
Atretic Systemic AV Valve	4 +
Univentricular Heart	2 +
Atrioventricular Septal Defect (Canal)	3

* All W 1 month of age
** Overlapping Groups
\+ Overlapping Groups
PDA = patent ductus arteriousus
ASD = atrial septal defect
VSD = ventricular septal defect
TGA = transposition great arteries

MANAGEMENT OF COARCTATION OF THE AORTA IN INFANTS

Patients with isolated coarctation presented with minimal congestive heart failure and all had preoperative LV hypertension (140-210 mmHg). Patients with associated defects presented with severe congestive heart failure and in 16 such patients prostaglandin (PGE) infusion was used preoperatively to maintain ductal patency. In our series, of 25 infants less than 1 year old, only 5 had isolated coarctation (23%), while the other 20 had associated anomalies (77%). The indication for operation was progressive congestive heart failure despite aggressive medical management. In all 25 patients with subclavian flap technique was used for repairing the coarctation. Eight patients underwent pulmonary artery banding, 4 at the time of coarctation repair, one 2 days before and 3 within two weeks of the repair of the coarctation. Banding was performed only when the intracardiac lesion was to be not amenable to primary definitive repair.

SURGICAL TECHNIQUE

The chest was entered through a lateral thoractomy via the fourth intercostal space on the side of the descending aorta. The pleura overlying the subclavian artery and proximal descending aorta was dissected as a flap and retracted anteriorly with clear identification and preservation of the vagus nerve and its recurrent laryngeal nerve. The distal aortic arch, subclavian artery, ductus (ligamentum arteriosium) and proximal descending aorta were dissected. See Figures 1–4. The ductal structure was doubly ligated in continuity. The subclavian artery was dissected to beyond the origin of the vertebral artery, which was ligated and transected. The subclavian artery was ligated distal to the vertebral artery. Clamps were placed on the aortic arch and a curved clamp

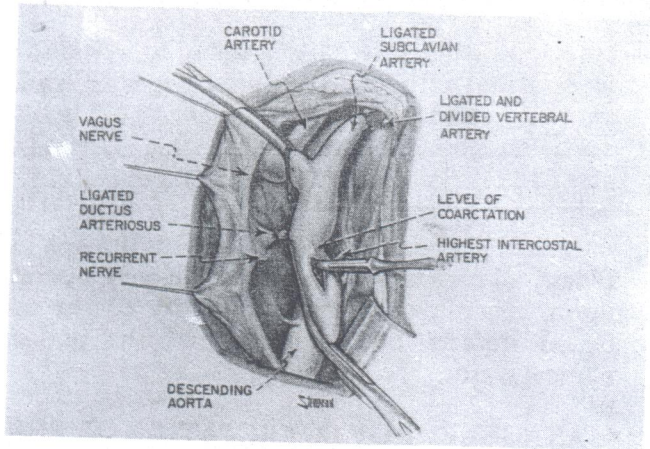

FIG. – 1.

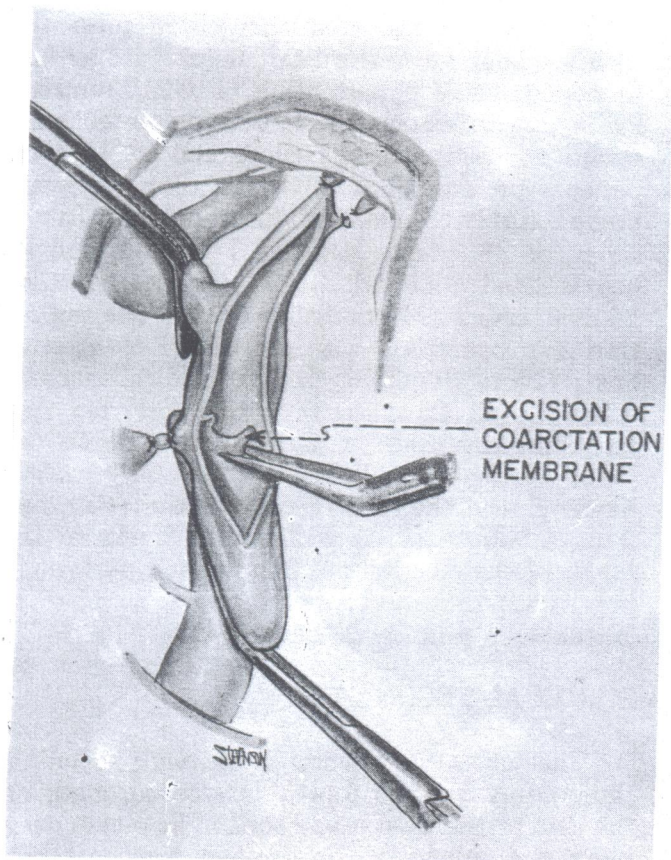

FIG. – 2.

on the descending thoracic aorta well below the level of the coarctation, even as low as the third intercostal artery. An incision was made in the aorta distal to the coarctation, through the coarctation, and carried along the length of the ligated subclavian artery. The coarctation membrane or posterior fibrous ridge was carefully resected and the transected subclavian artery brought down as

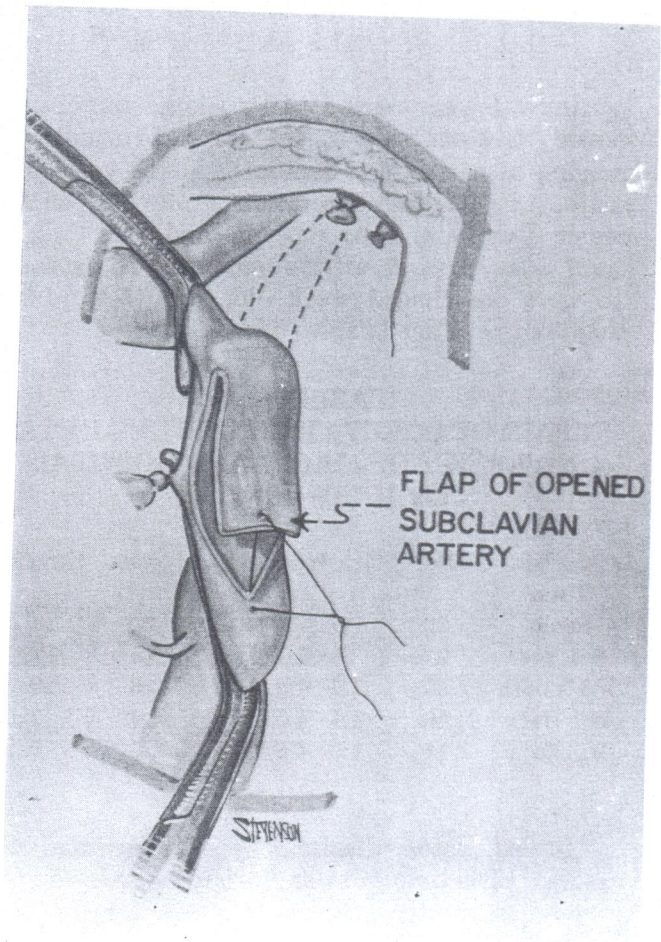

FIG. – 3.

a pedicled flap and sutured over and beyond the site of coarctation with 6—0 monofilament suture. Clamp time ranged from 8 to 30 minutes. The importance of extending the original incision well below the coarctation into normal aorta must be stressed, to maximize the width of the subclavian flap at the level of the coarctation, thereby avoiding the risk of "re-coarctation".

MANAGEMENT OF AORTIC COARCTATION IN CHILDREN

Children beyond the stage of infancy who have coarctation of the aorta present either with mild congestive heart failure or with upper extremity hypertension. Symptoms of headache or claudication my be variably present. Recently, we even saw a young woman who presented with recurrent (2 previous) spontaneous abortions; hypertension with each pregnancy had been incorrectly diagnosed as pre-eclampsia.

Repair of coarctation was done in 14 patients greater than 1 year of age. No other associated

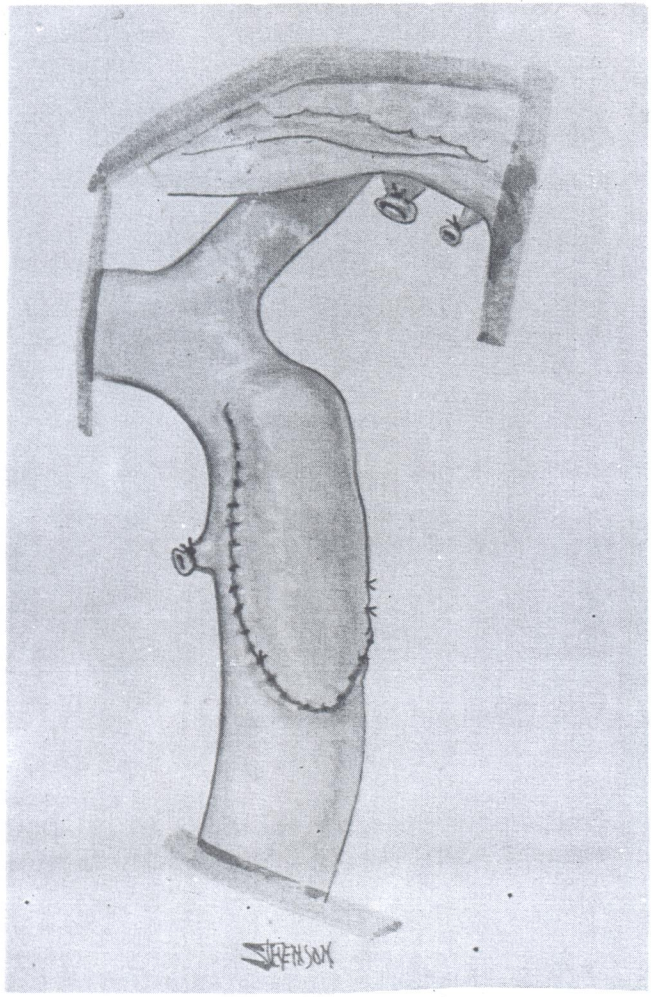

FIG. — 4.

Surgical Technique for subclavian flap aortoplasty for coarctation of aorta.

cardiovascular anomalies were present in this group of patients, except one patient who had an aberrant origin of the right subclavian artery. Five of the patients presented with recurrent coarctation. Four presented following resection with an end-to-end anastomosis and one after a subclavian flap repair elsewhere. Two of these were corrected with a Dacron patch angioplasty, 2 had a left subclavian artery to descending aorta graft, and in one a subclavian flap repair was found technically feasible and was therefore performed. Of the 9 older patients presenting with coarctation for the first time, three had resection with end-to-end anastomosis, 4 had subclavian flap aortoplasty and 2 had Dacron patch angioplasty. (Table - III).

TABLE — III
OPERATIONS FOR
COARCTATION OF AORTA

Subclavian Flap Repair	30
Dacron Patch Aortoplasty	4
Left Subclavian Artery to Descending Aorta Bypass Craft	3
Resection With End-to-End Anastomosis	3

RESULTS

Six of the 25 infants died. The only intraoperative death was among the 8 patients who had simultaneous pulmonary artery banding (Table - IV). One patient with a small VSD and severe mitral stenosis, acidotic and seizing preoperatively, died in a low output state 36 hours after repair and pulmonary artery banding. Another premature baby with mid-septal VSD's died from aspiration pneumonia and sepsis 2 weeks after coarctation and pulmonary artery banding. One newborn whose coarctation repaired at 2 days of age, underwent emergency open aortic valvulotomy at 5 days, had a smooth immediate postoperative course and died sud-

TABLE — IV
INDICATIONS FOR PULMONARY
ARTERY BANDING

Simultaneous	
Univentricular Heart/Systemic AV Atresia	2
Atrioventricular Septal Defect with Small Left Ventricle	1
Mid-Septal VSD (1900 gm)	1*
TGA/Tricuspid Atresia/VSD	1*
Mitral Atresia/Small LV/VSD (BAS)	1*
Delayed	
Multiple VSD's (2 wks, 3 mos)	2
TOTAL :	8

* Death

A-V = atrioventr
A-V = atrioventricular
LV = left ventricle
VSD = ventricular septal defect
TGA = transposition great arteries
BAS = ballon atrial septostomy

denly one day after the procedure (? potassium overdose). There were two late deaths, one 6 months after satisfactory coarctation repair in

TABLE – V
MORTALITY RELATED TO AGE AT OPERATION

Age at Operation	Patients Operated	Patient Died	% Age Mortality
W 1 Week	16	5	31%
W 1 Week	24	1*	4%

* Pulmonary Vascular Disease at 11 Months

TABLE – VI
MORTALITY RELATED TO ASSOCIATED DEFECTS

	Patients Operated	Patients Died	% Age Mortality
Isolated Coarctation	17	0	0
Coarctation with Associated Defects	23	6	25%

a baby with a hypoplastic left ventricle. Another 11 month old died following subsequent VSD closure and autopsy revealed grade IV pulmonary vascular disease (Tables V and VI).

Thirty-four survivors continue to do well up to 4 years postoperatively. All have normal weight gain and are normotensive. No patient has a measured arm-to-leg gradient greater than 10 mmHg. In one patient the VSD spontaneously closed, and in 2 other open VSD closure was uneventfully performed at 1 and 5 months of age. Another patient underwent a successful Mustard procedure at 3 months.

Nine patients have undergone repeat catheterization and none demonstreated any gradient, with satisfactory growth of the subclavian flap segment of repair (Table - VIII and Figs. 5 – 8).

DISCUSSION

Until a few years ago patients with coarctation of the aorta coming to surgery were older children and adults who were treated by the standard method of resection and end-to-end

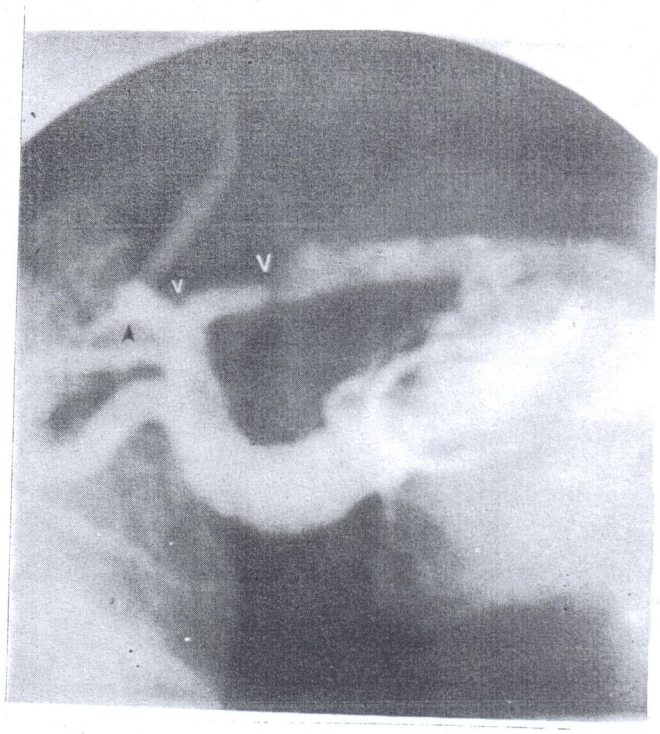

FIG. – 5.

Preoperative angiogram showing long coarctation segment distal to the left subclavian artery.

TABLE – VII
POSTOPERATIVE CATHETERIZATION FOR EVALUATION OF ASSOCIATED DEFECTS
9 Patients

Age at Op. (days)	Diagnosis	Age at Recath (months)	Gradient (mmHg)
2	VSD	1/30	0
2	TGA	3	0
3	Univentricle, PAB	6	0
3	VSD	1	0
3	Hypoplastic LV	6	5
3	Multiple VSD	3	0
10	TGA Tricuspid atresia, PAB	9	0
60	? Subaortic stenosis	8	0
90	Multiple VSD	28	0

anastomosis. With improvements in the care of the sick, premature newborns with multiple congenital defects, increasingly large numbers of

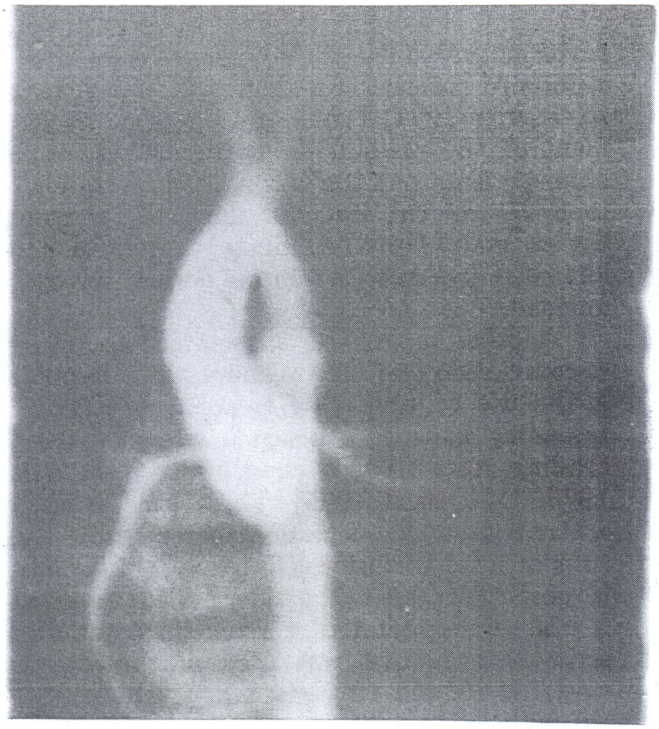

FIG. — 6.

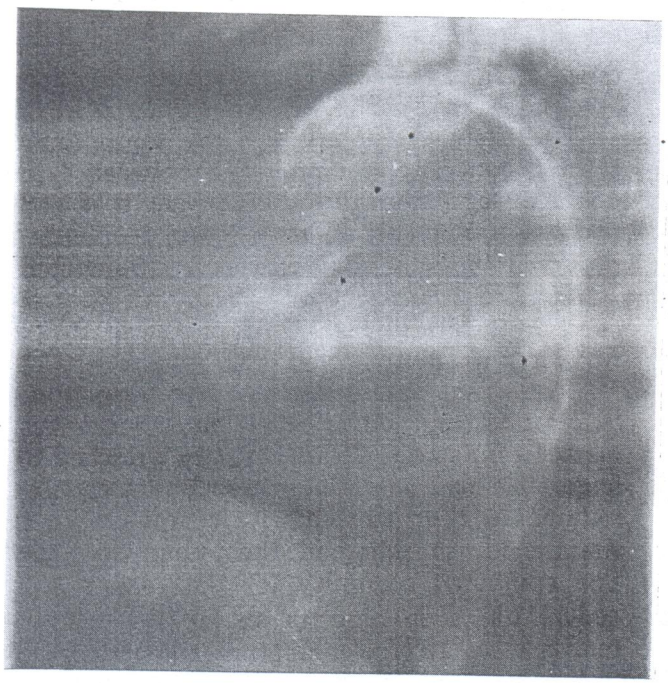

FIG. — 7.

AP and LAO views of postoperative anaiogram of patient shown in Fig. 5. Demonstrating restoration of the coarctation segment to normal Aortic Diameter by subclavian Flap Aortoplasty.

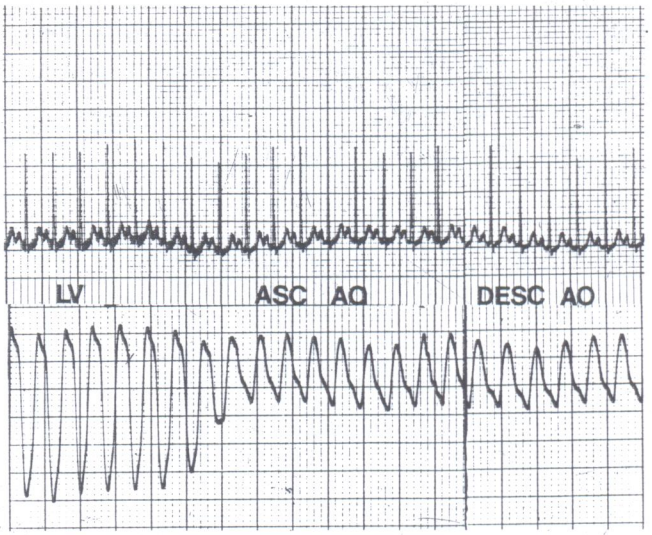

FIG. — 8.

Postoperative pressure tracing of patient in Fig. 5. Showing absence of pressure gradient between ascending and descending Aorta following subclavian Flap Aortoplasty.

patients with coarctation are presenting in the neonatal period. Although the coarctation is frequently only one of several cardiovascular anomalies in these patients, its correction often spells the difference between a baby who will survive and go on to repair of the remaining defects later and the one who will die of intractable congestive heart failure within the first few days of life.

We have adopted a very aggressive policy in the management of these extremely sick infants. While emergency surgical repair is essential to survival, the preoperative management is also very important. The latter includes a protocol of prostaglandin infusion, correction of acidosis, diuretics and inotropic support if necessary (dopamine, isoproteronol, but rarely digoxin). By maintaining patency of the ducts arteriosus and allowing distal perfusion, prostaglandin E_1 (PGE_1) infusions have dramatically reduced morbidity and mortality associated with surgical therapy.

Surgical repair of the coarctation can be done by various techniques. In the older child and adult, where the cross-sectional diameter of the aorta is already at or near maximum, the well-tried method of resection with end-to-end anastomosis is a satisfactory approach. We have used this method in only three patients in this series

(ages 14—16 years), with a very localized short coarctation segment and discrete membrane. Despite experimental evidence that the use of interrupted sutures for all or half of the anastomosis provided potential for normal luminal circumferential growth (Tawes, Aberdeen, and Berry, 1968), repair by this technique in infancy has resulted in a notriously high incidence of re-coarctation in most series, and is progressive with time (Williams et al, 1980).

In 1957 Vosschulte described the use of a prosthetic patch aortoplasty stressing the importance of excising the coarctation membrane and utilizing the base of the subclavian artery to assure an adequate lumen for growth. The theoretical advantages of this procedure are that it requires less aortic dissection, avoids sacrifice of the intercostal arteries, and avoids a circumferential suture line (Reul et al 1974, Fleming et al 1977, Sade et al 1979). It postulates some growth of the abnormal coarctation tissue. The problems of using a prosthetic material are inherent in this procedure, however, and late pseudoaneurysm formation, endocarditis and thrombosis have been reported (Olsson et al 1976, Kirch, Perry, Spooner 1977). A recent report by Bergdahl et al (1980) cites a 100% incidence of aneurysm formation in the aortic wall opposite the patch in long-term followup. We have used this procedure successfully in 4 patients with no complications t date, though we usually reserve it for re-operations.

The subclavian flap technique introduced by Walhausen and Nahrwold in 1960 has all the advantages of the prosthetic patch aortoplasty but in addition avoids prosthetic material and utilizes native tissue with a potential for growth. Sacrifice of the subclavian artery is well compensated by collaterals as has been observed in the decades of experience with the Blalock-Taussig shunt, and has been recently demonstrated in the long-term studies of arm growth following subclavian flap repair of coarctation by Todd et al (1983). However, caution must be exercised to ligate the vertebral artery to avoid a subclavian steal phenomenon (Saalouke et al 1978). Early followup of this procedure is very encouraging (Pierce et al 1978, Thibault et al 1975, Hamilton et al 1978), and long-term exercise testing results suggest this may be sustained (Fripp et al 1983).

All of the above described techniques require complete aortic occlusion. Though paralysis occurs in only 0.4% cases (Brewer et al 1972, Menasche et al 1978), the surgeon must be aware of possible ischemic damage if there is inadquate perfusion below the cross clamp. We know of no reports of paraplegia in neonates, but Hughes and Reemtsma (1971) have suggested routine measurement of the pressure in the distal aorta in the older patients — if the mean blood pressure is less than 50 mmHg, but the use of a temporary bypass graft) Gott heparinized shunt) or partial left heart bypass with femoral perfusion is recommended.

Another procedure, suggested especially for re-coarctation and long segment aortic hypoplasia is the use of prosthetic bypass grafts from the ascending aorta or subclavian artery to the descending aorta (Morris et al 1960, Weldon et al 1973, Edie et al 1975). These can be inserted with partial aortic occlusion thereby avoiding the risks of total occlusion. Obviously, this technique is most applicable in the older patient group since it utilizes a conduit of a fixed diameter. In our series this method was used in three patients aged 15, 16, and 19 years, two of whom presented with re-coarctation, with the third having unacceptably low distal aortic pressures on cross-clamping.

For the older child, the risk of surgery for coarctation of the aorta is generally 3—4% (Schusler and Gross 1962, Brom 1965, d'Allaines 1979). Elective surgery for the asymptomatic child is generally recommended between 4 to 6 years of age, though others argue that earlier correction may result in a lower incidence of post-coarctectomy hypertension (Shinebourne 1976, Liberthson et al 1979) or decreased coronary artery disease (Cokkinos et al 1979 and Rowen et al 1979).

The exact management of the neonate with coarctation and intracardiac defects remains a matter of debate. Classically, the presence of a large VSD and coarctation with congestive heart failure has been treated with coarctectomy and simultaneous banding of the pulmonary artery. The combined mortality for the two-stage procedure has been reported to be 50—62% (Macmanus et al 1977, Pennington et al 1979), contrasted with a 20% mortality for coarctectomy with subsequent definitive in-racardiac repair (Macmanus et al 1977). A recent report by Hammon et al (1982), however, cites no additional mortality if the VSD is closed and the pulmonary artery debanded within 6 months of the coarctation repair. Furthermore, Patel et al (1977), Pennington et al (1979), and Strafford et al (1980), all noted spontaneous closure

of the VSD in a significant proportion of patients following coarctation repair. Simultaneous correction of coarctation and associated intracardiac lesions has been reported in 6 infants without a death (Tiraboschi et al 1978) but this is not a standard procedure in most institutions.

Of our 8 patients with coarctation repair and pulmonary artery banding, 3 died, for a mortality of 38%. We had 6 patients with coarctation and VSD in whom coarctation repair alone was done as the initial procedure; all survived. In one patient the VSD closed spontaneously, two others have been successfully repaired at 1 and 5 months of age, one patient clinically well at 10 months of age and has not yet had the VSD repaired, and one patient whose coarctation was associated with critical AS and VSD underwent aortic valvulotomy and VSD repair 5 days after coarctectomy, did well initially and died suddenly one day thereafter. An 11 month old diagnosed as having only a VSD in earlier infancy, underwent uneventful coarctation repair and ductal ligation. She was re-admitted for closure of the VSD one month later, but died postoperatively. Grade IV pulmonary vascular changes were found. We have not yet performed simultaneous correction of coarctation and with associated intracardiac defects.

Based on our experience, therefore, we conclude that repair of coarctation of the aorta can be safely performed in any patient regardless of age or size. The mortality is that of any associated intracardiac anomalies. The subclavian flap technique offers reliably reproducible results and demonstrates satisfactory growth potential. This is especially useful in the infant and neonatal patients, while other procedures can be satisfactory utilized in older patients.

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