SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES AND TECHNOLOGY THIRUVANANTHAPURAM - 695011

PROJECT REPORT

NAME : DR. K. SIVAKUMAR
PROGRAMME : D.M. CARDIOLOGY
MONTH AND YEAR OF SUBMISSION : NOVEMBER 1999

PROJECT WORKS DONE

1. ANATOMIC SPECTRUM AND HEMODYNAMICS OF DOUBLE OUTLET RIGHT VENTRICLE - A DETAILED ANALYSIS OF 135 CASES WITH 2- DIMENSIONAL ECHOCARDIOGRAPHY AND ANGIOGRAPHY

2. LONG TERM FOLLOW-UP OF PATIENTS AFTER PERCUTANEOUS BALLOON DILATATION OF NATIVE COARCTATION OF AORTA – AN ANALYSIS OF 96 CASES

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CERTIFICATE

I, Dr K. SIVAKUMAR, hereby declare that I have actually carried out the projects under report.

Signature .....

Place : Trivandrum
Date : 10 Nov1999

Name in Capital letters

K. SIVAKUMAR

.....

Forwarded. He has carried out the two projects under report.

Signature

Head of the Department

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PROJECT REPORT
(PROJECT NO. 1)

TITLE

ANATOMIC SPECTRUM AND HEMODYNAMICS OF DOUBLE OUTLET RIGHT VENTRICLE - A DETAILED ANALYSIS OF 135 CASES WITH 2-DIMENSIONAL ECHOCARDIOGRAPHY AND ANGIOGRAPHY

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INTRODUCTION

Basically recognized by the origin of both great arteries from the morphological right ventricle, double outlet right ventricle (DORV) encompasses a variety of entities. This rare congenital anomaly has a frequency of 0.09 cases per 1000 live births.

HISTORY:

History of DORV dates back to 1898 when Vierodt described a partial transposition as what now would be called as DORV.(1) In 1949, Taussig and Bing described an anomaly with double outlet of right ventricle, large subpulmonic VSD, Aorta from RV and PA overriding the IVS. (2) In 1956, Witham classified partial transposition in to two groups, viz., Fallot type and Eisenmenger types. (3) In 1961, Neufeld classified DORV based on relation of great arteries (GA) and position of VSD.(4) These two criterion of classifying DORV was emphasized by subsequent workers Patrick and McGoon (5), Lev (6), and Zamora (7) as surgically important factors. In 1965, Carey and Edwards established angiographic diagnostic criteria for diagnosis of DORV (8):

(i) ***Both GA from right ventricle***

(ii) ***Aorta and Pulmonary artery arise at same horizontal plane***

(iii) ***A tongue shaped filling defect representing infundibular septum between outflow tracts***

(iv) ***Frequent malposition of aorta.*** In 1970, Hallerman added an additional criterion:(9)

(v) ***Mitral valve – semilunar valve fibrous discontinuity.***

NOMENCLATURE

Anderson defined (10) DORV as an abnormal ventriculo-arterial connection in which more than one half of circumference of both great arterial valves are connected to morphological RV. The corollary is that DORV and TOF are not mutually exclusive, and mitral – semilunar valve fibrous discontinuity is not essential for definition. The problem with this definition will be the difficulty in assessing override in surgery and autopsy.

Piccoli, Kirklin and Blackstone (11) defined DORV as almost similar to Anderson, but insisted that 90% of both GA should arise from RV. They added that Anderson's definition would include more anomalies in DORV group.

Van Praagh stressed that the diagnosis of TOF and DORV should be mutually be exclusive and emphasized subaortic and subpulmonic coni as diagnostic marker for DORV(12). Neufeld too emphasized need for semilunar valve – atrioventricular valve fibrous discontinuity for diagnosis (13). Donald Hagler (14) believed that cases with pronounced aortic override without aorto-mitral discontinuity would dilute the significance of the basic combination. Baron MG described bilateral infundibulum as a hallmark of DORV(15). Lev and Bharati oppose this contention and hold the same views as Anderson that "DORV can occur with semilunar – AV valve fibrous continuity". However Baron's views were considered to be of surgical importance since committing VSD to LV in aorto – mitral discontinuity is considered difficult. (15)

In many surgical and autopsy series of Norwood(16), Kirklin and Anderson(18), cases included as DORV even in the presence of aorto – mitral continuity. In Piccoli's series(11), Sridaromont (19), and Castenada (17), only DORV in the setting of atrioventricular concordance were included. ( ) Other anomalies as ***atrioventricular discordance, atresia of one atrioventricular valve, double inlet ventricle and atrial isomerism*** were excluded, since these anomalies alter the surgical strategies. Other series included these anomalies also since this group constitute a significant number in autopsy studies. Anderson quotes, " Each case of DORV warrants consideration on its own merits" (20).

Anderson (10) classified DORV as those with

(i) ***Normally related great arteries : spiralling relation of GA, line dissecting short axis of both GA is parallel to the IVS***

(ii) ***Side-by-side or D-malposed : Parallel relation of GA, line dissecting short axis of both GA is perpendicular to ventriculo infundibular fold***

(iii) ***L-malposed : Line dissecting both GA is parallel to IVS, Often the VSD is subaortic or doubly committed.***

Current classifications of DORV stresses on 2 morphological features namely site of ventricular septal defect and relation of great arteries. Based on site of VSD, Hagler (14) classifies DORV as

(i) ***Subaortic VSD***

(ii) ***Subpulmonic VSD***

(iii) ***Doubly committed VSD and***

(iv) ***Remote VSD.***

Zamora (7) made a new addition due to its surgical implications

(v) ***AV canal type or inflow VSD.***

Based on the relation of great arteries, 4 groups were recognised viz.,

(a) ***Normally related great arteries***

(b) ***Side – by – side great artery relationship***

(c) ***D – malposition of aorta***

(d) ***L – malposition of aorta***

Pathologic and angiographic reviews of Sridaromont (19), Lev and Bharati (6), and Zamora (7) allow categorisation of DORV based on these criteria. While Sridaromont defined cases with aorto – mitral discontinuity as DORV, others had less rigid definition. Here we report the morphological and hemodynamic features of 135 cases of DORV, including only patients with semilunar-atrioventricular valve fibrous discontinuity as specified by Van Praagh. This study excludes cases in whom more than half of both great arteries arise from morphological RV if aorto-mitral fibrous continuity is present.

AIM

This study aims at analyzing the anatomical spectrum of patients with double outlet right ventricle , the cardiac morphology assessed by detailed 2 – dimensional and doppler echocardiography and angiography in all cases and surgical findings in operated cases.

MATERIALS AND METHODS

A total of 160 patients with diagnosis of Double outlet morphological right ventricle (DORV) underwent cardiac catheterisation and hemodynamic study between 1978 and 1998, of which complete data of 135 patients were available for analysis. Diagnosis in each case was confirmed by detailed 2-dimensional echocardiography and angiography during cardiac catheterisation and/or surgery. This report highlights the clinical details, electrocardiographic and hemodynamic findings, description of cardiac morphology by combined echocardiographic and angiographic data.

In all the cases, clinical features as presence of cyanotic spells, history of congestive heart failure , history of central nervous system complications as cerebral infarcts or brain abscesses were noted. Clinical examination was done to assess the visceral and cardiac situs.

In chest X-ray, visceral situs, cardiothoracic ratio, pulmonary vascularity, side of aortic arch and other findings like features of juxtaposition of atrial appendages were noted. Electrocardiograms were examined for rhythm, P axis, PR interval, QRS axis, Right and Left ventricular forces , other abnormalities and arrhythmias.

Cardiac morphology was analysed in great details with combined informations from 2-D echocardiography and angiography. By echocardiography, viscero atrial situs, systemic and pulmonary venous drainage, size and site of ASD, morphology and function of atrio ventricular valves, Atrioventricular connections (concordant / discordant) , site, size and number of VSD, relation of great arteries, subvalvar, valvar and supravalvar obstructions of both great arteries, side and anomalies of aortic arch were noted.

From cardiac catheterisation, saturations in both great arteries, shunts, pressures in both great arteries and both ventricles, filling pressures, anomalies of venous drainage, juxtaposition of atrial appendages, presence and grade of atrioventricular valve regurgitation, ventricular morphology, atrio-ventricular concordance, size and site of VSD, location of VSD to great arteries, presence of additional VSD, relation of GA to each other , level of obstruction in outflow tracts (aortic and pulmonic), anomalies in great arteries and central pulmonary arteries, coronary anatomy, side of aortic arch were noted.

72 patients underwent surgical interventions till December 1998 and the surgical details of intracardiac morphology were also noted. The types of surgical interventions and their outcome were also analysed.

RESULTS

AGE:

The mean age of the 135 patients was 9.62 ± 8.14 years (mean ± one standard deviation). It ranged from 2 months to 46 years. 18 patients (13 %) were infants (Age less than one year). 48 patients (36%) were aged more than 10 years and 14 patients were aged more than 20 years (10 %).

TABLE I
AGE DISTRIBUTION

Age group	No of patients	Percentage of total
< 1 year	18	13%
1 – 5 years	41	31%
6 – 10 years	28	21%
11 – 15 years	20	15%
16 – 20 years	14	10%
> 20 years	14	10%

SEX:

There were 80 males and 55 females, the male : female ratio was 1.45 : 1.0.

CLINICAL PRESENTATION;

21 children (16%) presented with cyanotic spells. 20 patients (15%) had history suggestive of congestive heart failure and received digoxin and diuretics. 10 patients had central nervous system complications as infarcts or brain abscess. Abdominal situs were normal in 125 patients (93%) and inversus in 10 patients, liver dullness on left and gastric tympany on right (7%).

CHEST X-RAY FINDINGS:

Visceral situs was normal in 125 patients and inversus in 10 patients. The cardiac location was levocardia in 113 patients (83%), mesocardia in 5 patients (4%), dextrocardia in 17 patients (13%). The pulmonary vascularity was increased in 48 patients (36%), normal in 4 patients (3%) and reduced in 83 patients (61%). The aortic arch was right sided in 22 patients (16%), and left sided in 113 patients (84%).

ELECTROCARDIOGRAPHIC FINDINGS:

Sinus rhythm was noted in 128 patients (95%), junctional rhythm in 1 patient, one patient had complete congenital complete heart block with supra hisian narrow QRS escape, ectopic atrial rhythm in 5 patients. There was right axis deviation of

more than 200 msec. The mean frontal plane QRS axis was normal (0* to 90*)in 24 patients (18%), Right axis deviation (90* to 180*) in 53 patients (39 %), Left axis deviation (0 to –90*) in 24 patients (18%) and right upper quadrant axis (-90* to –180*) in 34 patients (25%).

While ECG criterion of right ventricular hypertrophy was present in all, 29 patients had features of left ventricular hypertrophy(21%). The arrhythmias noted included complete heart block in one, atrial flutter in one, paroxysmal narrow QRS tachycardia with no basal preexcitation in one, sick sinus syndrome with asymptomatic sinus pauses in one (Electrophysiological study confirmed sinus nodal dysfunction) Junctional tachycardia in one,Right bundle branch block (pre operative) in one patient.

MORPHOLOGY - VENO ATRIAL ANATOMY :

In the 10 patients (7%) with situs inversus, the IVC was left sided and abdominal aorta was right sided. IVC interruption with azygos continuation was noted in 8 patients (6%). Bilateral Superior vena cava (Persistent left SVC) was present in 23 patients (17%). No patient had viscero atrial discordance. 62 patients had no atrial septal communications (46%). Primum ASD was present in 9 patients (7%), Secundum ASD was present in 62 patients (46%), Single atrium was present in 2 patients. Anomalous pulmonary venous drainage was present in 5 patients (4%). One patient had PAPVC of left lower to coronary sinus, 2 had PAPVC of left pulmonary veins to left SVC, one had TAPVC to left SVC which was draining subsequently below to coronary sinus, one had TAPVC to right atrium. None of these anomalous venous channels were obstructed. Juxta position of atrial appendages were noted in 9 patients (7%).

TABLE II
ABNORMALITIES OF VENOUS DRAINAGE

Venous Anomalies	No of patients	Percentage
Systemic:		
Bilateral SVC	23	17%
IVC interruption	8	6%
Pulmonary:	(5)	4%
PAPVC to CS	1	
PAPVC to LSVC	2	
TAPVC to LSVC	1	
TAPVC to RA	1	
Normal:		
Normalvenous drainage	101	75%

MORPHOLOGY – VENTRICLES:

The ventricular relationship was normal in 114 patients(84 %), ventricular relations were anterior and posterior to each other (IVS profiled in coronal plane) in 8

patients (6%), side – by side relationship (IVS profiled in sagittal plane) of both ventricles in 13 patients (10%). All patients had ventricular septal defect as an outlet to left ventricle. None of the patients had intact IVS. 121 patients (90%) had single VSD and 14 patients (10%) had an additional muscular VSD. The VSD location was sub aortic in the majority, 69 out of 135 patients (51%), subpulmonic in 24 patients (18%), doubly committed in 2 patients, AV canal type of VSD in 29 patients (21%), remote or non committed in 11 patients (8%). The atrio ventricular relationship were discordant in 12 patients (9%).

TABLE III
LOCATION OF VSD IN 135 PATIENTS

Location of VSD	No of patients	Percentage
Subaortic	69	51
Subpulmonic	24	18
Doubly committed	2	2
AV canal/Inflow type	29	21
Remote	11	8

The ventricular septal defect was large and non restrictive in majority of the patients, 129 out of 135 patients (96 %). It was restrictive in 6 patients as assessed by doppler echocardiography and hemodynamics on cardiac catheterisation (4%).

The atrioventricular valve anomalies were noted in 25 patients (19%). Rastelli type C common atrioventricular valve was seen in 14 patients (10%). Mitral valve was hypoplastic in 4 patients, atresia of left AV valve was seen in 2 patients,Tricuspid atresia in one patient, hypoplasia of tricuspid valve in one patient, straddling tricuspid valve in 2 patients, supramitral ring and parachute mitral valve in one patient.

MORPHOLOGY – GREAT ARTERIES:

The relation of the great arteries to each other were normal in 11 patients where the aorta was right and posterior to pulmonary artery (8%), side by side relationship in 52 patients (39%), d-malposed relationship where aorta is right and anterior or directly anterior to pulmonary artery in 38 patients (28%), l-malposed relationship of great arteries where aorta is left of pulmonary artery, either anterior or to the side in 34 patients (25%). In no patient, aorta was left and posterior to pulmonary artery.

TABLE IV
RELATION OF GREAT ARTERIES TO EACH OTHER

Great artery relation	No of patients	Percentage
Normal	11	8%
Side by side	52	39%
D – malposed	38	28%
L – malposed	34	25%

96 patients had severe pulmonary outflow tract obstructions at various sites. All the remaining 39 patients had significant pulmonary hypertension. Other associations included persistent patency of ductus arteriosus in 23 patients (17%), major aorto pulmonary collaterals in 3 patients, sub aortic stenosis in 1 patient and post subclavian coarctation in one patient.

CORONARY ANATOMY:

The coronary anatomy was confirmed by aortic root angiography or ventricular angiography, and in operated patients with the help of surgical findings. The coronary origins were normal in 119 patients (88%). Single coronary artery from the right sinus was seen in 5 patients, single coronary artery from the left sinus in one patient, Right coronary artery from the left sinus and crossing the pulmonary outflow tract in one patient, left coronary artery from the right sinus in one patient, left circumflex from the right coronary artery and coursing behind the great arteries in one patient, right coronary artery from the left anterior descending coronary artery in one patient crossing anterior to the outflow tract, Left anterior descending coronary artery from the right coronary artery in 2 patients crossing the outflow, Left coronary artery to left pulmonary artery coronary cameral fistula in one patient, right coronary artery to right atrium coronary cameral fistula in one patient.

CATHETERISATION HEMODYNAMICS:

All patients underwent right and left heart study, oximetry runs and pressure recordings before angiographic studies to delineate morphology. Pulmonary arteries were entered in all patients with normal or elevated PA pressures. In patients in whom echocardiographic anatomy showed severe pulmonic stenosis, especially in the presence of systemic significant desaturation or history of hypercyanotic spells, PA was not entered and PA pressures were predicted from pulmonary vein wedge pressure tracings wherever obtained.

Pulmonary arteries were entered and oximetry sampled in 64 out of the 135 patients (47%). The mean PA saturation was 83.73 %. In the same group of 64 patients, the mean aortic saturation was 84.59 %. When all the 135 patients were included, the mean aortic saturation was 78.96 % and this group included those with severe PS, where PA were not entered.

52 patients (39%) had pulmonary hypertension defined as PA systolic pressures more than 30 mmHg . 42 patients (31%) had significant PAH with PA systolic pressures more than 50 mmHg. The mean PA pressure of the 99 patients (in whom a reliable PA pressure tracing could be obtained from either direct PA entry of indirectly though inference from pulmonary vein wedge pressure tracings) out of the total group of 135 patients (73%) was 45.73 mmHg. All the remaining 36 patients in whom the PA pressures could not be judged belonged to the severe PS subset and hence this mean PA

pressure value is an overestimate. The mean aortic pressures were 98.84 mmHg in the patient group.

SURGICAL PROCEDURES DONE

68 out of the total 135 patients (50%)underwent surgical procedures. 23 patients (17%)underwent intracardiac repair with or without transannular patch, one of which was a Senning's operation with pulmonary valvotomy for a patient with Taussig Bing anomaly. None of the patients underwent conduit surgeries. 12 patients (9%) had Glenn anastamosis, in which 11 patients underwent bidirectional Glenn shunt and one had classical Glenn anastamosis. 11 patients underwent Total Cavo Pulmonary Connection modification of Fontan surgery (8%). 22 patients (16%) had Blalock Taussig anastamosis, of which one had to be revised to a central aorto pulmonary shunt due to acute shunt thrombosis.

DISCUSSION

DORV has a wide spectrum of clinical presentations and morphological variations. We followed Van Praagh's definition of DORV by stressing on aorto mitral fibrous discontinuity for inclusion in this study. Patients with more than 50% aortic override without subaortic conus were not included. Sridaromont's series of 62 patients (19), Zhang's series of 92 patients (21) and Norwood's series of 57 patients (16) used similar diagnostic criteria. Anderson's group of 120 patients (18), were included with less rigid definitions.

CLINICAL FEATURES:

Clinical features suggested reduced pulmonary blood flow physiology in 83 of the total of 135 patients, of whom 21 patients had spells. This group accounts for 61.5% of the total cohort.

TABLE V
PROPORTION OF CASES WITH PULMONARY HYPERTENSION

Author	No of patients	% with PAH
Zhang et al (21)	92	13/92 (14%)
Anderson et al (18)	120	61/120 (51%)
Norwood et al (16)	57	35/57 (61%)
Sridaromont et al (19)	62	29/62 (47%)
Our series	135	52/135 (38.5%)

AGE:

The mean age of the cohort was 9.52 $\pm$ 8.14 years and ranged from 2 months to 46 years. Other series also mirror similar age ranges. Age of patients in Anderson's series ranged from 1 day to 44 years, Norwood's series ranged from 2 weeks to 20.4 years and Sridaromont's series ranged from 1 month to 33 years. Infants contributed to 13% (18/135) in our series in comparison with 11% (7/62) in Sridaramont's series and 32% (18/57) of Norwood's series. The age distribution in our group were highlighted in Table II.

SEX:

The male : female distribution was 1.45 : 1 in our cohort. In large surgical groups, they were 1 : 1 in Sridaromont's series(19), 57 : 43 in Anderson's series. But in a large Bohemian population study by Samanek et al , males predominated by 2.68 : 1 (22)

VISCERAL SITUS AND CARDIAC LOCATION:

We included all patients with abnormal situs and cardiac location in our group, since it is a morphological study. Sridaromont's (19) and Piccoli's (11) group

excluded patients with situs inversus, isolated dextrocardia and levocardia in their surgical series.

TABLE VI
VISCERAL SITUS AND CARDIAC LOCATION

Visceral situs	Cardiac location	No of patients	%
Situs solitus	Levocardia	114	85
Situs solitus	Dextrocardia	11	8
Situs inversus	Dextrocardia	6	4
Situs inversus	Levocardia	4	3

Van Praagh categorized spectrum of DORV malformations into 3 morphological groups of surgical relevance (12) :

(i) Malformations of conotruncus alone with associated VSD and subvalvar stenosis
(ii) Conotruncal anomalies associated with atrioventricular valve anomalies
(iii) Heterotaxy syndromes (abnormal cardiac and visceral locations)

Surgical series of Sridaromont (n = 62), Zhang (n = 92), Castenada (n = 73), Norwood (n = 57) did not include patients with abnormal visceral and cardiac locations and atrioventricular discordance with which they are closely associated.(19,21,16,17). In a significant number of patients with abnormal visceral and cardiac locations, definitive surgical procedures are precluded by associated multiplicity of anomalies like AV discordance, common AV valve, etc and hence excluded in major surgical series. Small reports of selected 19 patients with DORV and AV discordance by Batistessa et al (23)had 2 with situs inversus and 7 with dextrocardia. (9 out of 19 - 47%). 16 selected patients with DORV and common AV canal reported by Sridaromont et al had 6 patients (37 %) with isolated dextrocardia (24).

AORTIC ARCH:

Aortic arch was right sided in 22 out of 135 patients (16%). It carries little surgical significance except in palliative aorto pulmonary shunts.

ELECTROCARDIOGRAM:

Rhythm abnormalities were noted in 9 patients (7 %). Abnormalities included complete heart block, atrial flutter, paroxysmal SVT, junctional rhythm, ectopic atrial rhythm. 5 patients had ectopic atrial rhythm, superiorly directed P axis, of whom only one had IVC interruption. 7 other patients with IVC interruption had normal P axis. The only ECG report of DORV of 31 patients by Krongrad et al had 3 patients (10%) with rhythm abnormalities as atrial fibrillation, wandering pacemaker and ectopic atrial rhythm (25). Prolonged PR interval was found in 6 patients (4%), compared to 26% in Krongrad's series.

Mean QRS axis normal in 18%, RAD in 39 %, LAD in 18 %, and RUQ axis in 25%. Krongrad classified DORV in to two groups, Group I with no PS (Out of 12 , 7 had RAD, 3 had RUQ, 2 had LAD and none in normal axis) and Group II with PS

(Out of 19 patients, 16 had RAD, 1 had RUQ, 2 had normal axis). They concluded that predictive value of ECG was low(25). Our wide range of QRS axis variations are due to inclusion of all patients with visceral and cardiac situs anomalies and AV valve anomalies. Most patients in Sridaromont's selected group of patients with common AV canal had marked QRS axis abnormalities (24).

LVH defined in DORV as amplitude of QRS in lead V6 more than 15 mm in the presence of RVH (25) were noted in 29 patients (21%). Even though LVH is expected in LV volume overload conditions as

(a) Increased pulmonary blood flow without PS or pulmonary vascular disease
(b) Mitral regurgitation
(c) Restrictive VSD with suprasystemic LV systolic pressures (25), Korngrad could not identify correlates in 14 of 31 patients. 50% of patients in no PS group and 42 % of PS group had LVH.

VENOUS DRAINAGE ANOMALIES:

Anomalies of venous drainage noted in 34 patients (25 %), include bilateral SVC, IVC interruption, partial and total anomalous PV drainage). Normal systemic and pulmonary venous drainage were noted in 75 %. These anomalies and juxtaposition of atrial appendages (JAA) noted in 9 patients (7%) carry significance in surgical planning for Glenn and Fontan surgeries. Anderson's 120 patients (18)had venous anomalies in 4 % and JAA in 1 % , a selected group with situs solitus and AV concordance. Anomalies were more often seen in association with AV canal defects, 10 of 16 (63%) had persistent left SVC and 6 of 16 had (37%) anomalous pulmonary venous drainage(24) in Sridaromont's series of DORV with AV canal defect. 11% of Norwood's patients (16) had anomalies which included IVC interruption, left SVC and TAPVC. 10% of patients in Battistessa's group, comprising of a selected group of patients with DORV and AV discordance, had IVC interruption and TAPVC.(23) These surgically relevant venous drainage anomalies were less often found in Van Praagh's Group I patients comprising of isolated conotruncal abnormalities without AV valve and visceral situs abnormalities.(16)

ASD:

ASD was found in 53% of our cohort, 9 in primum location, 62 in secundum location and 2 had common atrium. Battistessa's group (23)had 68% and Sridaromont's group (19) had 25%. In a group of selected patients with persistent common AV canal by Sridaromont et al, 10 out of 16 patients had common atrium. Presence of ASD has significance only in few cases. Hagler noted ASD as a common association and the only eggress in patients with intact IVS and AV valve atresia. (14)

VSD SIZE:

Often VSD is large and nonrestrictive and presents free eggress for LV, Restrictive VSD causing a interventricular gradient in hemodynamic study of more then 20

mmHg constituted 6 cases in our group (4%) compared to 2% in Sridaromont's surgical series (19) and 17% of Hagler echocardiographic series(26). We did not come across any DORV with intact IVS. Few case reports of such cases exists (27).

VSD NUMBER:

14 patients in our group (10%) had additional muscular VSD in addition. In Sridaromont's series of 62 patients (19), 5% had additional VSD. In this study, 6 patients with subpulmonic VSD, 2 patients with AV canal VSD and 6 with subaortic VSD had additional muscular VSD.

VSD LOCATION:

The location of VSD was subaortic in 69 patients, subpulmonic in 24 patients, doubly committed in 2, AV canal type in 29 and remote to the origin of both great arteries in 11 out of the total of 135 patients. A comparison with other series is shown in the following table.

TABLE VII
LOCATION OF VSD IN VARIOUS SERIES

Author	Sridaromont	Anderson	Norwood	SCTIMST
	N = 60	N = 120	N = 57	N = 135
Subaortic	40/60 (67%)	60/120 (50%)	39/57 (68%)	69/135 (51%)
Subpulmonic	13/60 (22%)	39/120 (33%)	8/57 (14%)	24/135 (18%)
Doubly committed	2/60 (3%)	12/120 (10%)	2/57 (4%)	2/135 (2%)
Remote	5/60 (8%)	5/120 (4%)	1/57 (2%)	11/135 (8%)
AV canal type	N. A.	4/120 (3%)	7/57 (12%)	27/135 (21%)

A higher proportion of inflow or AV canal type of VSD was noted in our group. Since all the above mentioned surgical series had excluded patients with dextrocardia and visceral situs abnormalities, the proportion of cases with AV canal type of VSD is low. In our subgroup of 17 patients with dextrocardia, the location of the VSD was as follows:

(a) Subaortic in 6 patients
(b) Subpulmonic in 2 patients
(c) Remote in 1 patient
(d) AV canal type in 8 patients.

This may be one reason for a higher proportion of inlet (AV canal type) of VSD in our series.

In Sridaramont's exclusive analysis of a small cohort of patients with DORV with common AV canal,(24) out of 16 patients seen over 15 years, Dextrocardia was

noted in 6 patients (38%), a higher proportion than usual. In our study, out of 135 patients, 14 had common AV canal, akin to Sridaromont's group, of whom 5 patients had dextrocardia (3 of whom had situs inversus), and one patient had isolated levocardia with situs inversus. This again shows that AV canal type of VSD is more often found in abnormalities of visceral and cardiac situs.

VSD LOCATION AND RELATION OF GREAT ARTERIES:

The distribution of location of VSD in relation to the relation of great arteries to each other is shown in Table VIII:

TABLE VIII
RELATION OF VSD LOCATION TO GA RELATION

VSD location	Normal	Side by side	Dmalposed	L-malposed
Subaortic (n = 69)	10(8%)	33(24%)	17(13%)	9(7%)
Subpulmonic (n = 24)	1(1%)	6(4%)	8(6%)	9(7%)
Doubly committed (2)	-	1(1%)	-	1(1%)
Remote (n = 11)	-	7(5%)	2(1%)	2(1%)
AV canal (n = 29)	-	5(4%)	11(8%)	13(10%)

In comparison , Sridaramont's and Anderson's series had almost similar representation in each group. In Sridaromont's series of 60 cases, the various groups were as follows(19):

VSD location	Normal	Side by side	Dmalposed	L-malposed
Subaortic (n = 40)	2(3%)	28(47%)	9(15%)	1(2%)
Subpulmonic (n = 13)	-	4(7%)	6(10%)	3(5%)
Doubly committed (2)	-	2(3%)	-	-
Remote (n = 5)	-	5(8%)	-	-
No separate AV canal	group	na	na	na

In Anderson's series of 120 patients, the various groups were as follows(18):

VSD location	Normal	Side by side	Dmalposed	L-malposed
Subaortic (n = 60)	17(14%	32(27%)	4(3%)	7(6%)
Subpulmonic (n = 39)	-	15(13%)	24(20%)	-
Doubly committed (12)	6(5%)	5(4%)	1(1%)	-
Remote (n = 5)	-	3(3%)	1(1%)	1(1%)
AV canal (n = 4)	-	3(3%)	1(1%)	-

The minor differences in higher incidence of L- malposed aorta could be because of a considerable number of patients with AV discordance which has been included in the study group unlike the 2 author(18,19). In our 12 cases of DORV with AV discordance, the great artery relationship were:

a. Side – by – side in 2 out of 12
b. D – malposed in 1 out of 12
c. L – malposed in 9 out of 12

Yet another reason for a higher incidence of L – malposition of great arteries could be because of inclusion of all the cases with abnormal visceral and cardiac situs, which have been excluded in most of the surgical series. Our group of 135 patients included 17 with dextrocardia, of whom 6 had situs inversus and 11 had situs solitus. In this group, the great artery relationship were:

a. Side – by – side in 3 out of 17
b. D – malposed in 3 out of 17
c. L – malposed in 11 out of 17

AV VALVE ANOMALIES:

Morphological abnormalities of AV valves were noted in 37 out of the total 135 patients(27%). They are :

a.	Discordant AV valves	12	
b.	Common AV valve (Rastelli C)	14	
c.	Hypolastic mitral valve		4
d.	Atresia of left AV valve		2
e.	Tricuspid atresia		1
f.	Tricuspid valve hypoplasia	1	
g.	Straddling tricuspid valve	2	
h.	Supramitral ring + Parachute MV	1	

10 patients in Anderson's group (8%) had AV valve anomalies, which included 4 patients with common AV canal. However AV discordance were excluded in their group. Norwood's group had AV valve anomalies in 11 patients (19%), which included common AV valve in 7 patients, 2 with Ebstein's anomaly, one with hypoplasia of mitral valve (with straddling) and one with parachute mitral valve. Again, this group excluded AV discordance. Common AV canal in patients with DORV has been separately addressed by Sridaromont et al in 16 patients (24)and Freedom et al in 37 patients (29). Rigby in his editorial highlights the surgical significance of DORV with inflow VSD.

CORONARY ANOMALIES:

Our group witnessed anomalous coronaries in 16 (12%) patients, documented in either aortic root or ventriculography or in surgical notes. Lucia Gordillo et al described coronary patterns in 44 pathological specimens with DORV and found that the distribution of coronaries in those with normally related great arteries were more often

normal, in those with D- malposed great arteries, often resembled D-TGA. 20 % of coronary origins were abnormal in those with d- and l- malposed aorta(30). 25% of patients with DORV and common AV canal (24), 8 % of DORV with AV concordance and normal visceral situs in Sridaromont's series(19), reported coronary anomalies. In centers increasingly using RV to PA conduits, these anomalies may carry less significance compared to use of transannular patches.

OXIMETRY:

Pulmonary artery could be entered in only 64 patients (47%). In these patients, the mean PA saturation was 83.73% and the aortic saturation was 84.59 patients. This group included many patients with Taussig Bing anomaly and hence the PA saturation was much higher. When all the 135 patients were considered, the mean aortic saturation was 78.96%.

PRESSURES:

The PA pressures could be directly measured in 64 patients and indirectly through PV wedge pressures in 35 patients. The mean PA pressure in this group of 99 patients were 45.73 mmHg. The mean aortic systolic pressure in the entire cohort was 98.84 mmHg. The 36 patients in whom PAS pressure could not be assessed belonged to the severe PS by echocardiography, and hence the mean PA pressure is an overestimate. 52 patients had PAH (>30 mmHg), 42 had severe PAH (>50 mmHg).

HEMODYNAMICS OF SUBAORTIC VSD:

Of the 69 patients in this group, 27 had pulmonary hypertension defined as PA systolic pressures more than 30 mmHg. The remaining 42 patients had severe pulmonary stenosis. PA were entered in 32 of these 69 patients.

SUBAORTIC GROUP :OXIMETRY :

Aortic saturations were more than PA saturations in 25 of the 32 patients in whom PA were entered. The saturations were similar in 5 patients and PA was higher in 2 patients. The mean aortic saturation was 87.6% and the mean PA saturation was 81.6%. In comparison, Sridaromont's group of 40 patients with subaortic VSD, (19)had 24 with higher aortic saturations, 7 with similar saturations in both aorta and PA and 9 with higher PA saturations.

SURGICAL OUTCOME OF SUBAORTIC VSD: PAH GROUP

In this group, 3 patients had additional muscular VSD. Of the 27 patients with PA systolic pressures more than 30 mmHg, the mean PA pressure was 70.07 mmHg. The mean aortic pressures in this same group was 95.18 mmHg. Of these 27 patients, 10 underwent intracardiac repair, of whom 2 patients died in the post operative period. 9 patients were considered inoperable due to pulmonary vascular disease in 7 patients, hypoplastic left ventricle in one and restrictive VSD in one patient. One died before surgery and 7 await surgery with conduit.

SURGICAL OUTCOME OF SUBAORTIC VSD: PS GROUP

Of the 42 patients with severe pulmonary stenosis, 3 patients had an additional muscular VSD. 9 patients underwent intracardiac repair, one of whom died. 5 patients had BDG anastamosis . 6 patients had TCPC modification of Fontan surgery, of whom 2 died. 9 patients underwent BT shunt. One another patient who had a shunt thrombosis after BT shunt underwent a central aorto pulmonary anastamosis. 12 patients await intracardiac repair with conduit.

HEMODYNAMICS OF SUBPULMONIC VSD:

In this study, there were 24 patients with subpulmonic VSD. 11 patients (46%) had pulmonary hypertension defined as PA systolic pressures more than 30 mmHg. The other patients had severe pulmonary stenosis on echocardiogram, of whom PA was entered in only 2 patients. The proportion of patients in whom PA could be entered were 13 out of 24 (54%).

OXIMETRY IN SUBPULMONIC VSD:

The mean saturations in the PA of the 13 patients in whom PA could be entered was 94.6%. The aortic saturations in the same group was 82.4%. In 11 of the 13 cases, the PA saturations were more than aortic saturations. In 2 cases,the PA and aortic saturations were similar. In a similar analysis by Sridaromont (19), out of 13 cases of subpulmonic VSD, all patients had PA saturations more than aorta.

TAUSSIG – BING ANOMALY:

The PA pressures were higher in 11 out of 24 patients. In this group of patients, which come under the category of TAUSSIG - BING anomaly, the mean PA pressures were 83.45 mmHg. The mean aortic pressures in this group were 98.84 mmHg. In this group of 11 patients, the great artery relationship were normal in 1, side by side in 4, d-malposed in 3 and l-malposed in 3 patients. Of Sridaromont's 13 patients with similar morphology, 4 had side by side relation and 9 were d-malposed. In our group, 5 patients had additional muscular VSD also.

SURGERY IN SUBPULMONIC GROUP:

There were 11 patients with pulmonary hypertension and 13 patients with severe pulmonary stenosis. There were 5 deaths in this group, 3 before any surgery were done, one after an attempted intracardiac repair,
one after BDG anastamosis. One patient underwent Sennings surgery, VSD closure and pulmonary valvotomy, 3 underwent BDG anastamosis, 2 underwent TCPC modification of Fontan, 2 had Blalock Taussig Anastamosis, 2 are decided inoperable due to disconnected left pulmonary artery. The remaining patients are waiting for surgery.

INFLOW VSD GROUP ; HEMODYNAMICS:

There were 28 patients in this group. The PA was entered in 13 out of the 28 patients. 4 patients had pulmonary hypertension (PA sys pressure > 30 mmHg) . 24

patients had severe pulmonary stenosis. The mean PA pressures in the 4 patients were 87.75mmHg.

OXIMETRY: INFLOW VSD:

In this PAH group, the mean PA saturation was 86.5 mmHg, the mean aortic saturation was 78.25 mmHg. If the entire group of 13 patients in whom PA were entered were taken into account, the mean PA saturation were 85.7%, the mean aortic saturations were 83.7%. 4 patients had PA saturations more than aorta, 3 had PA saturations less than aorta, 6 had similar PA and aortic pressures.

SURGICAL OUTCOME: INFLOW VSD :

Morphologically, 12 patients of this group had Rastelli type C common atrio ventricular canal defect. Others had 2 separate AV valves. 11 patients in this group underwent surgery. Due to the following reasons, 10 patients were continued on medical followup. 4 had severe pulmonary hypertension and pulmonary vascular disease, 3 had unclear PA anatomy on echocardiography and angiography, 1 had PAPVC contraindicating a Fontan type of surgery, one had straddling tricusped valve with significant regurgitation, one was backed out after an attempted BDG anastamosis due to high normal PA pressures. 11 patients had surgery, one underwent intracardiac repair, 5 underwent BT shunt, 2 had BDG anastamosis, and 3 had TCPC surgery. 7 patients await surgery.

DOUBLY COMMITTED VSD:

In this group, we had only 2 patients. Both patients had PA saturations less than aortic saturations. One had pulmonary hypertension (PA systolic pressure was 75 mmHg). He underwent intracardiac repair. Other patient has severe pulmonary stenosis. This patient underwent BDG anastamosis.

REMOTE VSD:

11 patients in our group had VSD location unrelated to both great arteries. 4 patients had pulmonary stenosis and 7 patients had pulmonary hypertension. The mean PA pressures in the 7 patients were 80.42 mmHg.

REMOTE VSD – OXIMETRY:

In the 7 patients with pulmonary hypertension , PA could be entered. In the remaining 4 patients with severe pulmonary stenosis, PA were not entered. The mean PA saturation in this group were 83.6%, the mean aortic saturation being 88.1%. In 6 patients, the aortic saturations were more than PA and one had similar saturation. In none of the patients, PA saturations were more than aorta. In Sridaromont's series(19), of the 5 patients with remote VSD, the aortic saturations were higher in 3, PA was higher in one and both were similar in one.

REMOTE VSD – SURGICAL OUTCOME:

One patient underwent intracardiac repair, 2 patients with pulmonary stenosis had BT shunt (one of whom died after shunt thrombosis). All the other patients are on medical followup.

CONCLUSIONS

1. Double outlet right ventricle encompasses a variety of entities, with a varying spectrum of different morphologies and hemodynamic profiles.

2. Male to female ratio in this cohort was 1.45 : 1

3. 39% of patients had pulmonary hypertension, others had severe pulmonary stenosis

4. Abnormal visceral and cardiac situs were seen in 15% of cases

5. Abnormalities of pulmonary or systemic venous anomalies are present in one fourth of cases

6. 54% of patients had an atrial septal defect, often in secundum location

7. Common location of VSD in this group was in subaortic location, VSD in Inflow location were more often found in patients with visceral heterotaxy.

8. 10% of patients had additional VSD in muscular septum (a point of surgical significance)

9. Abnormalities in atrioventricular valves were noted in 27% of this group

10. Great arterial relationship was more often side by side, but in patients with visceral heterotaxy, atrio ventricular discordance, common AV valve, great arterial relations were d- or l- malposed.

11. 50% of patients in this cohort underwent surgical procedures

12. Complex anomalies like visceral heterotaxy, atrioventricular discordance, morphological anomalies of atrio ventricular valve, abnormal great arterial relationships precluded definitive biventricular repair in majority of patients.

13. Univentricular repair (Staged modifications of Fontan surgery) improved the clinical outcome in these patients with complex intracardiac anatomy

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PROJECT REPORT
(PROJECT NO. 2)

TITLE

LONG TERM FOLLOW-UP OF PATIENTS AFTER PERCUTANEOUS BALLOON DILATATION OF NATIVE COARCTATION OF AORTA – AN ANALYSIS OF 96 CASES

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INTRODUCTION

Coarctation of aorta (CoA) is a discrete stenosis in the proximal descending thoracic aorta, first described in 1760 by Morgagni at autopsy. The first surgical repair carried over by Crafoord in 1944 started a plethora of surgical techniques each vying each other in improving immediate and long-term results. (1) Trans-catheter dilatation was first described as a surgical alternative to surgery in 1982 (2). Despite many years of experience with numerous treatment modalities of this seemingly "simple" lesion, there continues to be a debate on what is best therapeutic approach for the pediatric and adult patient. The controversies are due to interpretation of immediate and medium term hemodynamic results, procedural risks, modification of natural history by treatment and personal bias(3).

CoA presents as a waist or concavity of the outer contour of aorta, opposite to PDA ligament with a variable degree of hypoplasia of isthmus and transverse arch, the latter influencing the long-term results of dilatation.(4,5). Histology shows a thickened avascular intimal tissue over a deformed and thickened media within the vessel wall. In infancy, ductal smooth muscle fibers extend to encircle the aortic wall and contribute to stenosis. (6). Depletion and disarray of the medial elastic tissues is also a feature though not consistent, a potential morphologic substrate for later aneurysm formation and dissections(3).

In the Natural history of CoA , 90% of patients with isolated unoperated CoA die before 50 years of age of heart failure, endocarditis, aortic rupture, intracranial hemorrhage, hypertension or associated valvular heart disease(7). Surgery favorably alters this as shown by 73% survival at 44 years by Brouwer (8), though surgically treated cohort remain a risk for late hypertension and premature death if operated beyond infancy(8,9). Recurrent CoA and higher early mortality due to associated lesions dominate surgery in infancy, Kappstein reporting 32% hospital mortality and 41% reCoA (9), reintervention for re CoA carrying a significant risk of mortality and morbidity(10,11)

Though Thomas Sos on a postmortem specimen in 1979(12) and Lock on surgically excised specimen showed feasibility of balloon dilatation(13), Singer reported the first successful dilatation of post-surgical reCoA in infancy and (2) and Lababidi in native CoA in infancy (15). Vascular injury following dilatation comprising of intimal and medial tears, some extending to adventitia have been described in detail from surgical specimens by Ho (16), Intravascular ultrasound (IVUS) by Sohn (17), Trans esophageal echocardiography (TEE) by Erbel (18), Angiography by Rao. Though

healing may result in a structurally weakened wall, its impact on long-term follow-up has not been well described. As a result of favorable blood flow patterns after dilatation and histologic healing, favourable remodeling of aortic contour, diameter of dilated CoA segment gradually increasing on follow-up and approaching isthmus dimensions in late restudy(19). Restenosis secondary to fibrointimal proliferation and ductal tissue too remains a possibility in infancy (20).

Optimal balloon size remains a point of no consensus, suggestions for selecting the balloon size equal to twice or thrice the CoA diameter (21), equal to the isthmus (22), equal to the size of aorta at the diaphragm (23), or 2 mm larger than isthmus (22) come from previous studies. The multi-centered Valvuloplasty and Angioplasty of Congenital anomalies registry (VACA) (24) showed lack of correlation of hemodynamic improvement or aneurysm formation with balloon size. This view is endorsed by Rao (25), and a large meta-analysis on 970 dilatations by McCrindle (26). Variable anatomy, aortic wallstructure, catheter techniques and balloon characteristics could explain these findings.

Conventional vascular access had been through femoral arteries though in some, umbilical arteries, antegrade approach across PDA, trans-septal puncture through venous route, through right heart after 1st stage Norwood surgery have been described in reports(3). Balloon dilatation has been uniformly effecting immediate hemodynamic success defined as post dilatation gradients of < 20 mmHg in 78 to 91 % of patients (22), including the large metaanalysis by McCrindle (26). Risk factors for suboptimal results included higher presystolic gradients, earlier procedure date and older patient date(26).

Restenosis defined as reoccurrence of gradient of > 20 mmHg after a successful procedure was described in 13% of patients overall (27), 60% in infancy and 7.3% in older children. Fletcher quoted a restenosis rate of 23%(22) and Rao described 27% (28) in their series. Risk factors for late restenosis included young age, isthmal hypoplasia, (Isthmic diameter < 2/3 of the ascending aorta proximal to origin of right innominate artery), narrow CoA segment < 3.5 mm before and < 6 mm after angioplasty(22). Kaine reported that z-Value for isthmus more than –1, age at angioplasty and associated anomalies had no effect on angioplasty outcome (29).

Incidence of aneurysms following dilatations vary due to varying definitions for the same, one followed often being an aortic wall contour deformation, with a diameter 1.5 times the aorta a diaphragm(19). Rao stressed the need to rule out a ductal bump, which gets outlined better after the procedure(30). MRI has been an excellent noninvasive method for morphologic study of arch and might help to determine presence of

aneurysms, degree of remodeling or reCoA.(3) Follow-up studies done with chest X-rays, angiography, MRI scans reveal an incidence of late aneurysms after balloon dilatations of 5 to 43%, the former values in the earlier reports (28). Discrepancies are not only related to the lack of consistent definition of aneurysm, but also to the lack of high-resolution angiogram both prior and following dilatation (3). Some technical factors contributing to these aneurysms have been prolonged inflations and use of longer balloons. Earlier apprehensions about risks of operating on coarctation complicated by aneurysms after dilatations have been allayed in subsequent studies by Minich (31).

Other complications include thromboembolic episodes in <2% (22), femoral occlusion in 10 to 16%, and rarely paradoxical hypertension. Though previously balloon dilatation was practiced in sick unstable infants, due to improved medical and surgical techniques, primary surgery would be preferred today (3). Acceptable indications for dilatations include peak to peak Coarctation gradient of >30 mmHg, brachial hypertension >95th centile and uncontrolled hypertension with heart failure. Arch morphology is an important factor to decide on method of treatment. Isthmus hypoplasia defined aortic isthmus < 2/3 of ascending aorta by Rao (25), < 70% of descending aorta by Fletcher (22) and isthmus z-value of < -2.16 by Kaine (29) have guided inclusions for balloon dilatations.

There is no current clear consensus as to the most appropriate candidate for dilatation. In native CoA in infancy, high incidence of vascular access complications, higher restenosis rates, common association of arch hypoplasia, may favour surgical treatment in view of improved outcome after surgery. Conte in his series of 151 infants operated at less than 3 months of age (32) found a 10 year actuarial survival rate of 98% in isolated CoA but higher rates of restenosis.

In older child, balloon dilatation has been a safe procedure with restenosis rates of 7 to 12 % from large studies.(22)and 5% incidence of aneurysms. In this group, the hazard of late death stems from hypertension and CAD, intracranial and aortic aneurysms, dissections and complications of aortic valve diseases. Even though surgical results in this age group are excellent with early mortality of 1%(33), restenosis of less than 5%, aneurysms still occur after surgical procedures , greater risks after patch aortoplasty, less after subclavian flap surgery and end to end anastamosis(34).

In adults with native CoA, Fawzy (35) and Tyagi (36) found restenosis in less than 10% of their cohorts, and in a meta analysis,

McCrindle noted success in 74% of the patients (87). In ReCoA after surgery, balloon remains the first choice in view of higher surgical risks. Periadventitial scarring may protect against excessive vessel damage(3). In reCoA after previous balloon dilatation, a distinction has to drawn between recurrence (reappearance of gradient after an initial success) and residual CoA (persistence of gradients during the initial procedure). While the former may be more amenable due to excessive fibrous tissue overgrowth, the later has to be analyzed for technical reasons (small balloon, inaccurate measurements), histologic reasons (ductal tissue, elastic recoil), and anatomical factors (isthmus diameter) before planning a repeat balloon dilatation.(3) With this background, we intend to analyse the long-term results of our patients who underwent dilatation of native coarctation of aorta.

AIM OF THE STUDY

1. To study the long-term outcome after balloon dilatation of native Coarctation of aorta
2. To analyze the morphological associations of mitral and aortic anomalies and shunts in this cohort of patients with coarctation of aorta.
3. To analyze the parameters influencing the immediate and late hemodynamic results after balloon dilatation of coarctation of aorta.

MATERIALS AND METHODS

A total of 96 patients with a diagnosis of coarctation of aorta underwent hemodynamic study and percutaneous balloon dilatation of coarctation between the year 1988 and 1998. These patients form the subjects of this study.

Diagnosis of each patient was confirmed after detailed clinical examination, chest skiagram, electrocardiogram, 2-Dimensional and Doppler echocardiogram. All patients underwent cardiac catheterisation and balloon dilatation, majority in the same sitting, 6 patients in a separate sitting. Left heart catheterization was performed from femoral access by the Seldinger's technique. All patients received 100 units per kilogram bodyweight of un-fractionated heparin intravenously after vascular access. The coarctation segment was crossed with straight tip guidewire through an end hole catheter (usually Goodale Lubin, Gensini, or right Judkins catheter). Aortic arch angiograms were performed in AP, LAO 60^0 cranial 20^0 and lateral views, wherever maximum delineation of arch anatomy is obtained, both pre and post angioplasty. The balloon size in all the instances were decided based on the isthmus diameter recorded on angiogram. All immediate complications including access complications, dissections were recorded. A successful dilatation procedure was defined as reduction of systolic peak gradient after the procedure to less than 20 mmHg.

Hemodynamic and angiographic restudy were performed in all patients on regular follow-up, who consented for the Restudy procedure. In the restudy the hemodynamic pullback gradients across the dilated site, presence of aneurysms were recorded. All the patients on regular follow-up, including those who did not consent for restudy were regularly followed up every 1-2 years for presence of systemic hypertension and clinical gradients across the coarctation by sphygmomanometric blood pressure recordings of upper and lower limbs.

The clinical outcome and procedural details of patients of 2 groups, one with an initial successful procedure (post dilatation gradient less than 20 mmHg) and other with initial inadequate result (post dilatation gradient more than 20 mmHg) were analysed using student t test for comparison of means of variables and chi-square test for comparison of proportions, and a p-value of less than 5% was considered statistically significant.

RESULTS

96 patients underwent balloon dilatation for coarctation of aorta between the years 1986 and 1998. These patients formed the subjects of this study. The mean age of the patients were 16.32 $\pm$ 10.21 years. There were 12 infants. 63 patients (66%) were older than 10 years and 41 (42%) were older than 20 years. Males predominated the study, male to female ratio was 72 : 24 (3:1).

All the patients in this cohort before balloon dilatation had systemic hypertension. Systemic hypertension was defined by 5th Joint National Committee recommendations on detection, evaluation and treatment of high blood pressure, 1993, for patients aged above 18 years and more than 95th centile in patients aged less than 18 years as recommended in 2nd task force report on blood pressure control in children, 1987 (37).

13 patients presented with congestive heart failure needing digitalisation and diuretics. Chest skiagram showed cardiac enlargement defined as cardiothoracic ratio more than 50% in 22 patients (23%). Rib notching was noted in 44 patients (46%). ECG showed features of Left ventricular hypertrophy in 46 patients (48%), LVH with strain pattern in 9 patients (9%), Right ventricular hypertrophy in 11 patients (11%), (All patients showing RVH were infants), Partial right bundle branch block pattern in 3 patients, Complete LBBB in one patient, Preexcitation (WPW syndrome) in one patients without any tachyarrhythmias, 1st degree AV block in one patient.

5 patients had secundum atrial septal defect, one had primum ASD, 5 had ventricular septal defects, 4 of which were small muscular and one was a large defect with pulmonary vascular disease and right to left shunt, patent ductus arteriosus in 4 patients with minimal shunts and one patient had both a small muscular VSD and small PDA. 79 patients (82%) had no intra or extra cardiac shunts. Of the 6 patients with ASD, 5 underwent surgery after a successful balloon dilatation, one female patient with a < 1.5:1 shunt remains on medical follow-up. Among the patients with VSD, due to negligible shunts, none were operated. One patient with large VSD with pulmonary vascular disease, was lost to continued follow-up after balloon dilatation. All patients with PDA had shunts detectable only on doppler echo, not detectable on follow-up. None were operated for PDA. 29 patients had pulmonary artery mean pressure more than 20 mmHg, 36 patients had pulmonary artery systolic pressure more than 30 mmHg.

Echocardiogram revealed mitral and aortic abnormalities and they are listed in Table I.

TABLE I
Mitral and Aortic valve Abnormalities

Abnormalities		Number of patients
Mitral valve:		
Cleft Anterior mitral leaflet		1
Congenital mitral stenosis		4
Parachute mitral valve		1
Mitral valve prolapse with leak		7
Rheumatic MR		1
Aortic valve:		
Bicuspid aortic valve		49
Subaortic membrane		4
Aortic regurgitation	Grade 2	5
	Grade 3	5
	Grade 4	1
Aortic stenosis	Mild	6
	Moderate	1
	Severe	8

Other anomalies detected included :

1. One 32 female patient with right ventricular endomyocardial fibrosis, who had successful dilatation, no heart failure or hypertension on follow-up.
2. Two patients with sub arachnoid hemorrhage secondary to anterior communicating artery aneurysms, one had balloon dilatation before aneurysm surgery, other following clipping.
3. Two patients developed ascending aortic aneurysms on follow-up with annulo aortic ectasia, with moderate aortic incompetence (both were non-Marfan).
4. Two patients with subaortic membrane with severe LVOT obstruction had simultaneous balloon dilatation of both subaortic membrane and coarctation.
5. One patient had rheumatic mitral regurgitation.

The hemodynamics during the balloon dilatation procedure is described in table II.

Table II
Immediate hemodynamics

Hemodynamic parameter	Mean ± S.D*
Ascending aortic systolic pressure	155 ± 32.6
Descending aortic systolic pressure	99.8 ± 21.5
Pre dilatation coarctation gradient	57 ± 23
Post dilatation coarctation gradient	21.2 ± 15.7
Reduction of systolic gradient	35.1 ± 19.5

*All values in mmHg

The anatomy of the aortic arch showed discrete postsubclavian coarctation in 89 patients, presubclavian in 3 patients of whom 2 were post surgical (jump graft patients , hypoplasia of aortic arch in 2 patients, sigmoid coarctation in one patient. The mean balloon diameter was 10.78 ± 3.23 mm.

In this cohort, 92 patients had dilatation of native coarctation and 4 procedures were done on post surgical jump graft patients on the native coarct segment for residual gradients. 39 patients had inadequate result with a residual gradient of more than 20 mm Hg out of 96 patients (41%). 55 patients had initial procedural success (59%) defined as a postdilatation gradient of less than 20 mmHg.

A total of 47 patients out of 96 patients underwent hemodynamic restudy and angiography at an average of 1.85 years. The systolic gradient across the coarctation segment was 25 ± 21 mmHg. 21 patients had a gradient of more than 20 mmHg. 15 out of this 47 patients had systemic hypertension and were receiving antihypertensive drugs. 32 patients had no hypertension. After the restudy, 7 patients in this group had surgical repair for coarctation.

Of the 96 patients, 82 patients had regular follow-up and alive at last review (86%). The mean follow-up duration was 6.32 ± 3.76 years. 52

patients had a follow-up of more than 5 years and the longest follow-up was for 12 years. 3 infants (3%) had a late death, 2 to 6 months after the procedure. 11 patients were lost for follow-up(11%). At the last follow-up, the mean clinical systolic gradient across the coarctation was 18 $\pm$ 15 mmHg.

The follow-up details of the 39 patients who had initial inadequate result (post dilatation gradient > 20 mmHg) are given in Table III

Table III
Follow-up of 39 patients who had inadequate result

Restudy done (n = 22)		No restudy done (n = 17)	
Gradient >20 mmHg (n = 11)	Gradient $\leq$ 20 mmHg (n = 8)	Lost for follow-up (n = 4)	Regular follow-up (n = 13)
4 – surgery 3 – redilated 3 – no HTN 1– post surg	None had HTN No increase in gradient on follow-up	1 – died (infant) 3 – no follow-up	3 – surgery 3 – no HTN 3–fixed HTN, no grad 4– HTN, grad

The long-term follow-up of the 57 patients who had successful dilatation defined as a post dilatation gradient of less than 20 mmHg is given in table IV.

Table IV
Follow-up of 57 patients who had successful dilatation

Regular follow-up (n = 47)		No follow-up (n =10)
No systemic hypertension on follow-up (n = 33)	Systemic hypertension (n = 14)	8 patients lost for follow-up
Gradient > 20 mmHg 7 patients	Gradient > 20 mmHg 7 patients	2 infants died after a period of 2 – 6 months
Gradient ≤ 20 mmHg 26 patients	Gradient ≤ 20 mmHg 7 patients (fixed HTN)	

In this group of patients who had successful dilatation, hemodynamic restudy was done in 25 patients, 22 patients had no hemodynamic restudy, 9 patients lost to follow-up. The restudy details are given in table V

Table V
Hemodynamic profile of patients on regular follow-up who had initial successful dilatation

Restudy done (n = 25)				No restudy done (n = 22)			
Systemic HTN (n = 7)		No HTN (n = 18)		Systemic HTN (n = 7)		No HTN (n = 15)	
Grad > 20 mmHg	Grad ≤ 20 mmHg	Grad > 20 mmHg	Grad ≤ 20 mmHg	Grad > 20 mmHg	Grad ≤ 20 mmHg	Grad > 20 mmHg	Grad ≤ 20 mmHg
N = 3	N = 4	N = 3	N = 15	N = 4	N = 3	N = 4	N = 11

The differences between the patient characteristics, initial hemodynamics, procedural variables, restudy hemodynamics, follow-up gradients and residual hypertension were compared in the 2 groups of patients who had initial successful and initial inadequate dilatations (postdilatation gradient less than and more than 20 mmHg) in the table VI.

The mean age of the patients in the two groups Group I with initial procedural success and Group II with initial inadequate result were 14.8 and 18.6 respectively. There were 7 infants in group I and 5 in group II. This was not statistically significant. Similarly the male:female ratio were not statistically different in the two groups.

Among the hemodynamic variables, a severe coarctation represented by higher ascending aortic systolic pressure and higher peak systolic gradients were indicative of initial inadequate result and the difference was statistically significant. Other variables as descending aortic pressures, balloon size were not statistically significantly significant in the 2 groups.

On follow-up, the clinical gradient across the coarctation measured by sphygmomanometry and cardiac catheterisation restudy were not statistically significant in the 2 groups. Similarly both groups had similar incidences of residual hypertension and need for repeat balloon dilatation. However more patients with initial inadequate results had required surgical correction of coarctation, and the difference was statistically significant.

Table VI

Parameter	Initial Successful Result Gradient ≤ 20 mmHg	Initial inadequate result Gradient > 20 mmHg	t value	P
Patient characters:				
No of patients (%)	57(59%)	39(41%)	-	-
Age in years	14.8 ± 11.3	18.6 ± 10.4	1.67	NS
No of infants (%)	6 (11%)	4 (10%)	-	NS*
Sex ratio	2.4 : 1	4.6 : 1	-	NS*
Males in %	70%	82%	-	NS*
Procedural Details:				
Asc Ao sys pr(mmHg)	147.6 ±30.4	165.0 ± 33.0	2.66	Signif
Des Ao sys pr (mmHg)	99.4 ± 22.2	96.3 ± 20.6	0.69	NS
Gradient	48.2 ± 17.1	68.7 + 24.2	4.87	Signif
Balloon size (mm)	11.0 ± 3.08	10.5 ± 2.94	0.37	NS
Post gradient	10.9 ± 6.2	35.5 ± 13.4	-	-
Restudy:				
No of patients restudied	26 (46%)	21 (54%)		NS*
Gradient on restudy	23.0 ± 17.9	29.3 ± 25.5	1.42	NS
Follow-up:				
Years of follow-up	6.1	6.7	-	NS*
No lost to followup	10 (17.6%)	4 (10%)	-	NS*
On regular follow-up	47 (82.4%)	35 (90%)	-	NS*
Follow-up gradient	17.7 ± 15.3	20.8 ± 15.1	0.98	NS
No HTN on follow-up	70%	54%	-	NS*
HTN on follow-up	30%	46%	-	NS*
HTN with gradient	15%	17%	-	NS*
Fixed HTN; no gradient	15%	29%	-	NS*
Reprocedure:				
Balloon redilatation	4 (7%)	3 (8%)	-	NS*
Surgical repair	3 (5%)	8 (21%)	-	Signif

- * p value > 0.05 (Chi square test)
- t value with degrees of freedom = 94 (Paired t test)

Procedural complications are listed in table 6.

Table 6
Procedural complications

Complications	No of patients	Follow up
Femoral artery thrombosis	8	6 –Thrombolysis with streptokinase 2 – conservative Rx
Femoral AV fistula	1	Spontaneous thrombosis
Pseudo aneurysms	11	2 patients operated 9 patients non progressive aneurysm
Mediastinal hematoma	1	Explored, no aortic tear visualised
Dissections	4	2 patients operated 2 not present on follow-up restudy
Dye allergy	1	Conservative Rx
Post dilatation hypertensive crisis	1	Medical Rx with SNP, ACEI
Transient visual defect	1	CT scan negative, no deficit on follow-up

11 patients out of the total of 96 patients underwent surgery on follow-up. 4 patients underwent dacron patch aortoplasty, 4 had end to end anastamosis after resection of the coarctation segment, 2 had interposition graft, 1 had jump graft. 3 patients (all infants) died on follow-up after 2 to 6 months. 82 patients (85%) had eventfree and surgery free survival at last review (mean follow-up period of 6.3 years).

DISCUSSION

Our study presents the longterm follow-up of patients who had balloon dilatation of native coarctation of aorta. A previous report of initial immediate hemodynamic benefits of the procedure of 46 earlier patients of this cohort was presented by Subramanyan et al in 1992(38). The two variables found to have significant influence on the immediate results have been identified in the study as balloon size and isthmus size in relation to transverse arch diameter and descending aorta on univariate analysis and only balloon size on multivariate analysis. This study present the long term follow-up of 96 consecutive patients who underwent balloon dilatation and includes the initial 46 patients also.

Procedural Success:

Procedural success have been variably defined in literature. Lock defined success as coarct diameter more than 30% and peak gradient reduction of more than 50 mmHg (39). He stressed on the fallacies of gradients as confounded by blood flow across coarctation site and blood flow across collaterals, diameter of coarctation segment, vascular resistance at vasculature above and below the coarctation segments and heart rate. But since none of these parameters could be assessed in routine catheterisation procedure, gradients remain to be the indicator of identifying results. In our study, we used post dilatation gradient of less than 20 mmHg to indicate good result. This cutoff was used in previous studies by Fawzy et al (23), Fletcher et al (22) and others. Rao et al define good result as less than 20 mmHg and fair result as upto 30 mmHg (28). Fletcher opined that post gradient of 20 mmHg is a good divider in the absence of large patent ductus arteriosus or very large collaterals(22). In our study, which comprises a heterogenous population of various ages and different morphology, procedural success was achieved in 59% of the study group.

Among the various parameters, only ascending aortic systolic pressures before dilatation and pre dilatation peak gradient across the coarctation segment (related variables) were predictive of inadequate initial result. The other variables including age of patient, sex, balloon size (which in our cohort was selected based on isthmic size) were not predictive of initial procedural success.

While 25% of patients who had initial procedural success developed gradients of more than 20 mmHg on follow-up, 28% of patients who had initial inadequate results had a reduction of gradients to less than 20 mmHg

on follow-up. Hence the differences between the proportion of patients with residual gradients and proportion of patients with residual hypertension were statistically not significant when patients with initial procedural success were compared to those with inadequate results.

Age:

The mean age of the study group was 16.32 ± 10.21 years and comprised 12 infants. The mean age of group of Erbel et al was 27.3 (18), Tyagi et al was 19.6(36), Kulkarni et al was 17.8 (40) and Fawzy et al was 23.0 (23). Infants contributed to 25% of Fletcher's group (22). The age of patient had no impact on immediate outcome. Infancy has been found to be an important contributor to restenosis, especially in less than 3 months of age.(28) Age also plays an important factor in incidence of procedural complications relating to vacular access.

Associations:

Associated shunt lesions in this cohort were present in 18% of this cohort. This has varied in literature depending on the age group of the study group. In the group of Rao et al comprising predominantly of children of mean age of 3.9 years, 50 % of patients had associated lesions including complex anomalies.(28) Fletcher et al reported a 25% incidence of similar anomalies.(22)

Abnormalities of mitral valve were present in 14 patients in this group. Bicuspid aortic valve, a common association of coarctation was present in almost half of the patients. Abnormalities of left ventricular outflow tract including sub aortic membrane and valvar aortic stenosis and incompetence were found in 17 patients.

Follow-up gradients:

The method of assessing residual gradient in follow-up of patients in the study group had been with cardiac catheterisation in the 47 patients and clinical upper and lower limb blood pressure assessment by sphygmomanometry in all the patients. The latter has been the widely used method in majority of the studies. The lack of accuracy of doppler echocardiography in detecting the gradient has been studied by Rao et al, and found to lack correlation with catheterisation hemodynamics (41). In the study, he found correlation between catheterisation gradients and doppler peak velocity only after incorporating a regression equation, in which 2 other parameters, acceleration time and antegrade flow time were included. (41) Rao et al suggest to use doppler echocardiography in follow-up by recording

the initial post procedure descending aortic velocity and comparing the same with follow-up records in predicting recoarctation occurrence.

Among the 39 patients who had initial procedural inadequate result, on follow-up, 11 patients had a gradient less than 20 mmHg (28%). 8 of these 11 patients had their gradients shown by restudy and 3 clinically. This favorable hemodynamic phenomenon has been described as due to remodeling of aortic contour following histologic healing and normalisation of aortic flow after dilatation by Rao and Carey from angiogram done 6 to 30 months after the procedure (42). Magnetic resonance imaging studies by Weber et al also illustrates this phenomenon of favorable remodeling (43). Quantitative angiographic studies of Suarez de Lazo (44) and routine angiographic studies by Beekman (19) documented the diameter of dilated coarctation segment to isthmus ratio increasing with time and approaching unity in follow-up studies.

Recoarctation defined as recurrence of coarctation gradient more than 20 mmHg on restudy has been reported in all previous studies with an incidence of 9 to 90% depending on the cohort. In this study group, among the 57 patients who had successful dilatation, there was a recurrence of gradient of more than 20 mmHg in 14 patients (25%). Fawzy reported a low incidence of 9 % in his adult cohort (23). Rao in a large group of children (mean age 3.9 years) reported an incidence of 31% on follow-up (28). Redington quoted an incidence of 90% in his cohort of neonates (45). Rao et al have found that the best predictors of recoarctations to be age (infancy), pre and post dimensions of the dilatation segments, size of transverse arch and isthmus. In an infant with a high risk of recurrence of coarctation after the procedure, the decision for palliative angioplasty should be based on local surgical mortality and morbidity (3).

Differing definitions of restenosis also account for varying prevalence of recoarctations. Beckman defined it as a gradient of more than 50 mmHg.(19) Rao et al in their initial reports (28), defined it as gradient more than 30 mmHg or coarct diameter less than 50% of immediate post dilatation values.

Residual hypertension:

The prevalence of residual hypertension at the last follow-up in this series was 31%. The risk of persistent hypertension was not different in the 2 groups. While 17 patients among the 30 who had hypertension had clinical gradients of more than 20 mmHg, the rest had fixed hypertension with clinical

gradients less than 20 mmHg. Prevalence of residual hypertension has varied from 68% in Fawzy et al group (23) and 23% in Fletcher et al (22) group. This will depend on the mean age of the study group since early intervention is likely to prevent residual fixed hypertension.

Aneurysms:

Development of pseudo aneurysms after balloon dilatation remains the Achilles heel of the procedure. 11 patients (12%) had evidence of pseudo aneurysms at the dilated site on restudy. While 2 patients were operated for residual gradients , the rest continue to remain on medical follow-up, 5 of them have been documented not to increase in size after a repeat study. One patient who had evidence of mediastinal hemorrhage after the procedure underwent emergent thoracotomy uneventfully.

Rao et al reported 5% incidence of anuerysms in their follow-up study,(28) Fawzy reported a 9% incidence (23), Fletcher et al 2% (22), and in the large valvuloplasty and angioplasty for congenital anomalies registry (VACA registry), Tynan reported an incidence of 55% (24). The varying numbers are to a large extent contributed by the differing definitions, VACA registry defined aneurysm as a wall contour deformation around the coarct segment 1.5 times that of descending aorta at diaphragm (24). Rao stressed on the need for high quality angiography pre and post dilatation to diagnose aneurysms and need to exclude a ductal bump which might simulate a localised pseudoaneurym (28).

Subramanian et al postulated balloon size and isthmic hypoplasia as predisposers for aneurysms (38), though it has been negated in VACA registry (24) and Rao et al reports (28). Some of the procedural variables now considered to play a role include prolonged inflations, misinterpretation of isthmus size in selection of balloon and catheter/guide wire manipulations across the dilated site(3). Isner et al suggest that 1 – 2 year follow-up is inadequate since aneurysms develop upto 5 years after surgical procedure like patch aortoplasty (46).

The lack of knowledge of the natural history of these nonprogressive pseudo aneurysms have hindered widespread advocacy for balloon dilatation. Pinzon in a large study of angiographic restudy of post surgical patients, equally divided between end to end anastamosis, subclavian flap procedure and onlay patch aortoplasty, reported an incidence of 27 % after end to end anastamosis, 32% after subclavian flap surgery and 35% after patch aortoplasty (34). The criticism for their study was that only 27% patients in

their cohort of surgical patients were included in their angiographic study and they might inflate the proportions.

Isner et al reported that one of the key determinants of post procedure aneurysms is the inherent connective tissue anomaly of aorta (46). They reported a high incidence of cystic medial necrosis at autopsy and surgical excision specimens. Ho and Anderson negate this argument by showing similar incidence of cystic medial necrosis and normal histology in a number of normal and coarctation patients (4). Interestingly, we found 2 patients with large ascending ascending aortic aneurysms with annulo aortic ectasia with no features of Marfan's syndrome on follow-up.

Even though dissection are reported in only a small minority of patients in our group only (4 out of 96), the mechanism of dilatation is by producing intimo-medial dissections (3). They are often not detected after angiography, or may appear as irregularities of aortic contour. TEE and IVUS studies have been more accurate in detecting dissections with greater sensitivity than angiography. IVUS definitions for a minor dissection has been thin mobile membrane extending into the wall over not more than one-fourth of aortic circumference. All other extensive lesions are called major dissections. Follow-up studies with IVUS have shown partial complete healing (17). Among our 4 patients, 2 had no evidence of flaps on restudy, 2 were operated after the procedure.

Vascular access:

In all our patients, we used femoral arterial access. 8 patients (9%) had femoral arterial thrombosis with loss of femoral pulses, 6 of them thrombolysed with streptokinase and 2 with low molecular weight dextran and heparin infusion. All patients recovered pulses at the time of discharge. None of these patients at follow-up had any difference of blood pressure between the 2 lower limbs. Fletcher reported a 16% incidence of femoral arterial thrombosis (22), Rao et al reported 19% incidence in their pediatric cohort (28), Kulkarni reported a higher incidence of 38% (40), one third needing surgical thrombectomy.

The incidence has been declining due to usage of full doses of heparin during the procedure, usage of smaller profile balloons and widespread usage of arterial sheaths for the procedure and immediate removal of arterial sheaths after the procedure (3). The important patient variable has been patient age, younger infants carrying a high risk. Rao et al advise umbilical and transseptal access to obviate these problems (28). A disturbing feature in a

well conducted follow-up angiographic study which included iliac angiography in all pediatric patients,Rao et al report an incidence of late complete or partial stenosis of femoral artery of 14% (28).

Post dilatation hypertensive crisis:

The two distinct advantages proposed for balloon dilatation over surgery (3) has been avoidance of spinal cord complications and rebound hypertensive crisis (Post coarctectomy syndrome). A forme fruste of post coarctectomy syndrome characterised by occurrence of hypertensive crisis after balloon dilatation has been reported in 4 % of pediatric cohort of Rao et al (28). We witnessed such a phenomenon lasting 48 hours requiring sodium nitroprusside infusions and ACE inhibitors along with diuretics. This patient after 8 years of follow-up has no residual hypertension and no clinical gradient.

Reintervention:

Among our 96 patients, 7 patients required redilatation with balloon and 11 patients (11%) required surgical interventions. This accounts for 19% of the study group. While redilatation rates were similar in patients with initial procedural success and patients with inadequate initial results, surgery was needed in a significantly larger number of patients after an inadequate result (P=0.046). While recoarctations (initial procedural success) caused by excessive fibrous overgrowth might be amenable for repeat dilatation, residual coarctation (initial inadequate result) caused by recoil, isthmic hypoplasia, ductal tissue might not give procedural success. However if the initial dilatation has been inadequate due to selection of a smaller balloon or due to misinterpretation of angiographic dimensions in selecting the balloon size, repeat balloon dilatation might prove useful (3).

Comparison with surgical results:

In our study, we did not make any comparisons with surgical results. Shaddy (47) randomised patients to balloon dilatations and surgical procedures and reported similar mortality, less morbidity and complications and dramatic reductions in complications as paraplegia and paradoxical hypertension. Rao in their nonrandomised comparisons (28) reported similar conclusions.

Hanley in an editorial (14) stated that coarctation represented a continuum of disorders from discrete stenosis to tubular isthmus and arch hypoplasia, and surgical and percutaneous interventions should not be compared at random, but on a case – to – case basis. He stressed on the

occasional limitations of having 20 mmHg as a gold standard of successful procedure in the presence of large collaterals and also highlighted on the limitations of the different methods of assessing gradients : clinical sphygmomanometry, doppler echocardiography, and catheterisation. Studies of differing periods also should not be compared in view of the declining surgical morbidity in recent times.

CONCLUSIONS

1. The outcome of patients with post subclavian coarctation of aorta after balloon dilatation remains good on longterm follow-up. Only 11 % of patients required surgery. Balloon dilatation gives a surgery free survival for 89% of patients in this group.
2. Among the variables influencing an initial procedural success, age of the patient, sex, size of the balloon, (which in our cohort was selected based on angiographic measurement of isthmic size) were not predictors. Initial higher predilatation gradients and predilatation ascending aortic pressures were indicative of less adequate results.
3. Restenosis rates in this study is 25%, patients need regular follow-up to assess need for reintervention.
4. 28% of patients with initial post dilatation gradients more than 20 mmHg have reduction of gradients on follow-up, indicating that favourable remodeling of aorta secondary to normalisation of flow may play a role. However a rigorous quantitative angiographic study is needed to confirm this finding.
5. Incidence of pseudo aneurysms after the procedure was 12% in this cohort. The lack of information about the long term natural history of these apparently non progressive aneurysms is disturbing, and needs consideration in patients who develop systemic hypertension on follow-up and female patients in future pregnancies.
6. Vascular access complications can be minimised by heparinisation, proper selection of hardware and immediate measures on detection of loss of pulses after the procedure.
7. The late occurrence of aortic dilatations and aneurysm formation in these patients , and the question of whether they are secondary to cystic medial necrosis needs to be studied.

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