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1. TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION A CLINICAL AND HEMODYNAMIC STUDY

2. PERIPHERAL PULMONARY STENOSIS

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CERTIFICATE

I, Dr. **KADER MUNEER. P** hereby declare that I have actually, performed all the procedures listed/carried out the project, under report.

Signature ..... 7/11/96 .....

Place:

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Capital letters

Date:

Forwarded. He has carried out the minimum requirement of procedures/etc.

Jayaraman 7.11.96

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PROJECT REPORT

TITLE OF THE PROJECT: **TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION -A CLINICA AND HEMODYNAMIC STUDY**

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CONTENTS

TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

A CLINICAL AND HEMODYNAMIC STUDY

INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart disease with an incidence of 0.2 to 2% of all congenital cardical anomalies[1]. In this anomaly the pulmonary veins have no connection with the left atrium and they connect to the right atrium or any systemic veins. TAPVC has a high mortality in symptomatic infants if left untreated. Prognosis is worse in children with pulmonary venous obstrctuion[1].

In about one third of patients, TAPVC is associated with major cardiac malformation[1]. Numerous classifications have been advanced for TAPVC. The most widely used and followed in this review is that by Darling[2]. Type I - anomalous connection at Supracardiac level, Type II - cardiac, Type III - infra diaphragmatic, Type IV - mixed, each may be obstructive or non obstructive. Here we report the clinical, hemodynamic and angiographic features of 108 cases of TAPVC.

MATERIALS AND METHODS

Between December 1976 to March 1996 a total of 108 patients with TAPVC were seen in our hospital. The diagnosis was made by ECG, X-ray, Echocardiographic and hemodynamic studies. 104 patients underwent cardiac catherisation and angiography. 61 patients were operated till March 1996 and the follow up ranged from 2 months to 55 months (mean 18 months).

RESULTS

There were 61 males and 47 females and their age ranged from 1 month to 32 years with a mean age of 6± 6.2 years. 36 patients were infants less than 1 year, 51 were between 1 year and 12 years and 21 were more than 12 years.

The patients were classified according to the mode of connection of Pulmonary venous channel to systemic veins (Group I to IV - Darling) (Table 1). 64 (59%) had the supracardiac TAPVC with a left sided vertical vein in 58 patients and right sided vertical vein in 6 patients.

33 (30%) had intra cardiac TAPVC which included 20 patients with pulmonary veins draining into coronary sinus and 13 with a direct drainage to right atrium.

9 patients (8.3%) had mixed form of TAPVC and in all of them the left upper pulmonary vein was draining into the left vertical vein and all other pulmonary veins were draining into the coronary sinus.

Infra diaphragmatic TAPVC was seen in only 2 patients (2%). One infant died before surgery because of pulmonary venous obstruction and severe pulmonary hypertension.

The other patient, an 8 year old girl with pulmonary veins draining to hepatic vein had obstruction in the distal common pul vein.

Pulmonary venous obstruction and subsequent pulmonary hypertension was seen in 17 patients (16%). Obstruction was taken as significant if the total pulmonary venous gradient was >10mm of Hg or gradient at the site of obstruction >4mm. All of these patients had significant pulmonary hypertension (P.A systolic pressure > 50 mm of Hg).

Of 17 patients with obstruction 11 had supracardiac TAPVC. Obstruction was at the junction of common chamber and vertical vein or between vertical vein and innominate vein. Pulmonary venous gradient could be measured in only two of the six patients with right ventrical vein. In one right ventrical vein was obstructive at it's opening to superior vena cava.

In 3 patients with TAPVC into coronary sinus, the obstruction was at the junction of pulmonary veins and coronary sinus in 2 and inside the coronary sinus in one. None of the intracardiac TAPVC with direct drainage in to right atrium were obstructive.

2 patients with the mixed type of TAPVC had obstruction of the pulmonary vein draining to coronary sinus. Both patients with the infradiaphragmatic type were obstructive.

Presence of obstruction and obstructive gradients were analysed by entering the venous channel during catheterisation and taking pull back tracings. In 3 patients the venous obstruction was diagnosed indirectly by the disparity between elevated P.A wedge pressure and right atrial pressure. Pulmonary venous gradient was measured in 45 out of 104 catheterised patients. In patients with obstruction (17/45) the total gradient ie from pulmonary vein to right atrium varied from 9 to 28 mm of Hg (mean - 15mm of Hg) and gradient at the site of obstruction was 4 to 13 mm of Hg (mean - 6 mm of Hg). In patients without significant obstruction the total gradients measured was < 10 mm of Hg. Inter atrial septum (IAS) was restrictive in one and he had balloon atrial septostomy.

97 patients (90%) had pulmonary hypertension (pulmonary systolic pressure > 30 mm of Hg). 39 of these patients had moderate to severe pulmonary hypertension (PAH) (PA systolic pressure > 50 mm of Hg). PAH was more frequent in infradiaphrgmetic type (100%) and supracardiac with right sided vertical vein (5/6).

We compared the pulmonary arterial saturation (PASat) with aortic saturation (Ao Sat) in 99 patients. The difference in saturation was considered significant if it was more than 5%. No significant difference between PA and Aortic saturation was seen in 63 of the 99 patients (64%). P.A saturation was more than aortic saturation in 26 patients and less than aortic saturation in 10 patients. Only 6 out of 59 patients with supracardiac TAPVC had PA saturation higher than systemic saturation compared to 9/18 (50%) of the coronary sinus type and 6/9 (66%) of the mixed type.

Most of the patients had significant left to right shunt. Average ratio of pulmonary to systemic flow (QP/Qs) was 3.5 in all types except intra cardiac variety were the shunt was 2.9:1.

Associated anomaly apart from ASD was noted in 25 patients. This included ventricular septal defect in 1, patent ductus arteriosus in 6, pulmonary stenosis in 11, peripheral pulmonary stenosis in 3, and single atrium, single ventricle, transposition and isolated levocardia one each. Two patients had pulmonary atresia.

Operative Management

61/108 (56%) patients underwent corrective surgery. 2 patients with pulmonary atresia underwent a palliative B.T. shunt. Corrective surgery included anastomsing the venous channel to left atrium or rerouting the veins to left atrium. In patients with mixed TAPVC the left upperpulmonary vein was left alone and only those draining to coronary sinus were rerouted. 13 patients died in immediate post operative period. Cause of death was residual obstruction in 5, Poor Rv function in 2, pulmonary Edema in 1, congestive heart failure in 1 and unknown in four. 10 out of 13 deaths occurred during the initial period of study.

Autopsy was available in 3 post operative patients. One had residual pulmonary venous obstruction. Myocardial necrosis was noted in one and the third patient had inferior vena caval drainage to left atrium.

3 patients had residual venous obstruction following surgery which was diagnosed by echocardiography in two and by cardiac catheterisation is one.

Out of 13 patients who died post operatively 5 were less than 1 year, 7 were between 1 and 12 year and 1 was more than

12 years. 5 patients died before surgery or catheterisation and all of them were infants.

Re catheterisation was done in 12 post operative patients and one patient had residual venous obstruction as suggested by elevated PA wedge pressure and normal left ventricular end diastolic pressure. Remaining 11 patients were free of any residual shunt or obstruction, and all of them had significant fall in P.A. pressure. Atrial arrythmia occurred in three post operative patients in the early post operative days. Two reverted to sinus rythm subsequently and one remained in junctional rythm.

DISCUSSION

Total anomalous pulmonary venous drainage is embryologically due to the failure of development of the common pulmonary vein and subsequent persistance and enlargement of the embryonic collaterals between the lungs and systemic veins. This condition was first described by Wilson in 1798[3].

The clinical manifestations of TAPVC depend on the presence and severity of pulmonary venous obstruction. Patients with pulmonary venous obstruction or restrictive ASD have early onset of symptoms and high mortality in infancy[1].

There is no sex preponderance in this study. One third of patients were infants. This is in contrast to other series were 70-80% were less than 1 year. The low prevalence of infants in this study may be due to lack of recognition of this condition and high mortality during infancy.

Supra cardiac TAPVC was the commonest type in our series as in other series[5,6,7]. The incidence of other types like right sided vertical vein, mixed TAPVC, intra cardiac types were comparable to other large series. But the incidence of infradiapharmatic type is rare in our series (2%) compared to

10% to 50% in the western series [5,6]. The low incidence of this type reflects the high infartile mortality because of the invariable pulmonary venous obstruction.

Pulmonary venous obstruction was seen in 16% of our patients. The reported incidence varies from 10% to 50%[7,8] in other large series. Pulmonary-venous obstruction was observed in both patients with infradiaphargmatic drainage. The obstruction was more frequent in the supra cardiac variety. In intra cardiac type only those with drainage to coronary sinus showed obstruction none of those with direct drainage in to RA were obstructive.

In this study, we analysed the gradients in the venous channel in obstructive type with significant pulmonary hypertension. It was found that total gradient between pulmonary vein and right atrium was more than 9 mm of Hg in all types with obstruction, compared to less than 10 mm of Hg in non-obstructive group. Hence a total gradient of more than 10 mm of Hg is highly suggestive of obstruction. At a single site, a gradient of more than 4 mm of Hg was seen only in obstructed type of TAPVC.

Most of the patients in our series had pulmonary hypertension. But significant PAH was seen in only 39/97 (40%) of patients. Mild PAH is due to the increased pulmonary blood flow with hyperkinetic pulmonary circulation. Pulmonary venous obstruction or rarely associated PDA or VSD is the cause of significant PAH.

Usually in TAPVC the oxygen saturation in all four cardiac chambers will be nearly equal, because of good mixing of blood at atrial level. 95 patients in our series showed nearly equal (<5% difference) systemic and pulmonary saturation. Higher pulmonary saturation was rare in supra cardiac type but more common in TAPVC with coronary sinus drainage due to selective streaming of the pulmonary venous blood to right ventricle.

Surgical follow up

61 patients have been operated till now, with 13 deaths. Most of these occurred in the initial period of study and equally common in infants and older children.

Residual pulmonary venous obstruction was the most common cause of early mortality. Most of the surviving operated patients were in functional class I and only 3 developed

significant symptoms due to residual pulmonary venous obstruction. The surgical morbidity and mortality in infants were comparable to some large series[9] where residual obstruction was the commonest cause of death.

Atrial arrythmias are known to develop after TAPVC or atrial septal defect correction. Only 3 patients developed atrial arrythmia post operatively in our series and two reverted to sinus rythm subsequently.

CONCLUSION

TAPVC is a rare congenital heart disease. Early diagnosis and treatment is important because of the high mortality in infancy[2] especially in the infra diaphargmatic type of TAPVC.

As TAPVC is an isolated anomaly in most cases, it is possible to have a successful correction followed by a normal life span[10]. The disease should be suspected in patients with features of large ASD with cyanosis or congestive heart failure. Those with obstruction to pulmonary veins will have severe PAH and respiratory distress in infancy. Long term follow up of operated patients showed no significant residual functional limitation[10].

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TABLE I

TABLE SHOWING DISTRIBUTION OF CASES ACCORDING TO DARLING'S CLASSIFICATION

Group	Number (%)	P.A saturation ≤5% of Aorta	Obstructed (Number)	PAH >50 mm of Hg	QP/Q (avg
Supra Cardiac					
-Left vertical vein	58(53%)	44	10	16	3.5
-Right Vertical vein	6(55%)	5	1	5	1.5
Intra cardiac					
-Coronary sinus	20(18.5	6	3	8	3.5
-Right atrium	13(12%)	5	0	5	2.9
Infradia-phragmatic	2(1.8%)	1	2	2	
Mixed	9(8.3%)	2	2	3	3.5

TABLE II

TABLE SHOWING COMPARISON OF INCIDENCE OF VARIOUS TYPES OF TAPVC FROM MAJOR STUDIES

Type of TAPVC	Burrough & Edward (7)	Delisle Et al (6)	Goswamy Et al (11)	SCT
Supra Cardiac	47	41	63	64
Intra Cardiac	31	26	27	33
Mixed	7	5	5	9
Sub-diaphragmatic	13	24	5	2
Others	2	4	-	-
Total	113	93	110	108

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PERIPHERAL PULMONARY ARTERY STENOSIS

INTRODUCTION

Stenosis of pulmonary arteries isolated or in association other cardiac defect are not uncommon. The incidence of this anomaly been reported to be between 2 and 3% of all congenital heart diseas The stenosis may be single or multiple. In two thirds of cases o associated cardiac anomalies are present. Valvular pulmonary steno ventricular septal defect, atrial septal defect and patent du arteriosus are the most frequent associated anomalies. Periph pulmonary stenosis (P.S) is also frequently seen with tetralog fallot[1].

In the present study we analysed 29 patients with periph pulmonary stenosis, excluding those with cyanotic congenital h disease.

Gay et al classified peripheral P.S. into four types. Type isolated constriction of main pulmonary artery or right or left branches. Type 2 is stenosis involving the bifurcation. Type multiple peripheral stenosis and Type 4 is a combination of main peripheral stenosis[3].

MATERIALS AND METHODS

Between 1976 and March 1996 we had 29 patients with peripl P.S. who underwent cardiac cathetirsation in our hospital. diagnosis was suspected clinically and later confirmed hemodynamic and angiographic studies. Those patients associated cardiac anomalies or isolated severe proximal ste were subjected to surgical correction and those with valvar had balloon dilatation of pulmonary valve. Re cathetirisatio done in five patients to assess the progression of the sten Others were followed up with regular echo cardiogr examination.

RESULTS

There were 15 males and 14 females and their age ranged from 10 months to 38 years. 15 patients were between 1 and 12 years. 12 were more than 12 years and 2 were less than 1 year.

2 patients had congenital rubella syndrome, one had Noonan syndrome and one had William syndrome. Majority of patients (23/29) were asymptomatic. 6 patients were in class II and none in class III or class IV. All patients with class II symptoms had either severe valvar PS or atrial septal defect as associated anomaly.

Cyanosis was seen in four patients. 2 had associated ventricular septal defect and two had severe multiple stenosis with right to left shunt at atrial level.

Second heart sound was widely split in four patients with associated valvar P.S. Continuous murmur was audible in 3 patients.

Chest x-ray showed cardiomegaly in five patients and pulmonary oligaemia in four patients. Plethora was seen in two patients, one with associated atrial septal defect and

another with ventricular septal defect. Electrocardiography showed right ventricular hypertrophy and right axis deviation in 8 patients. 3 Out of these 8 had associated valvular P.S.

Morphology of peripheral P.S. was classified according to angiographic appearance (Gay classification). 12 patients had either isolated supra vulvar obstruction or isolated RPA or LPA proximal stenosis (Type I). 13 had stenosis at the bifurcation (Type II). Two had multiple peripheral P.S. (Type III). and two had combination of above morphology (Type IV). (Table I)

20 patients (69%) had associated anomalies. Most common being vulvular P.S. (10). Patent ductus in 4, atrial septal defect in 3 and ventricular septal defect in 3. (Table II)

4 patients with severe valvular P.S. underwent successful balloon dilatation of pulmonary valve. Two patients had dysplastic value including one with noonan syndrome.

Severely elevated right ventricular systolic pressure was seen in 10 patients. This include 4 with valvular P.S., 2 with supra valvular membrane, 2 with atrial septal defect and 2 with severe multiple peripheral stenosis.

Repeat cardiac catheterisation was done in 5 patients, 2 had increase in the severity of peripheral stenosis following balloon pulmonary valvotomy and ASD closure. Other three had no significant change in gradient on repeat cardiac catheterisation.

DISCUSSION

Peripheral pulmonary stenosis is not a rare congenital malformation. The pathogenesis of this entity is unknown. Pulmonary artery and it's branches develop from three separate vascular components main pulmonary artery develops from Truncus arteriosus by partition with aortopulmonary septum, the proximal segment of RPA and LPA develop from sixth branchial arch on either side, the distal pulmonary artery branches derive from post branchial pulmonary vascular plexus. Any teratogenic insult on the components of the developing pulmonary arteries, may arrest their development, leading to atresia or stenosis. At least one teratogenic agent, `rubella virus' has been implicated in the pathogenesis of peripheral pulmonary stenosis.

In our study we had two cases of congenital rubella, one Noonan syndrome and one William syndrome with associated peripheral pulmonary stenosis.

Most of the patients with peripheral P.S. are asymptomatic and significant symptoms occur if the stenosis is severe and multiple or due to associated cardiac anomalies.

Most of the patients in our series with isolated peripheral P.S. were asymptomatic. Cyanosis is rare in peripheral P.S: two of our patients had multiple peripheral stenosis and cyanosis due to right to left shunt at atrial level[2].

Second heart sound may be normal or widely split, with normal or increased intensity. Four patients in our study had wide split second heart sound. In very proximal stenosis the pulmonary artery pressure proximal to obstruction is same as that of right ventricle and pulmonary valve remains open as long as the gradient between right ventricle and distal pulmonary artery persists.

Continuous murmurs are unusual in peripheral P.S. unless there is central pulmonary hypertension due bilateral stenosis or associated patent ductus arteriosus[8]. Usually peripheral P.S results in systolic murmur. Continuous murmur was noted in three of our patients.

Electrocardiogram usually shows normal or right axis deviation with features of right ventricular hypertrophy. Left axis deviation may be seen in patients with congenital rubella or Noonan syndrome. ECG in most of our patients showed normal or rightward axis. One patient had left axis deviation.

In patients with mild to moderate peripheral pulmonary stenosis the heart size and vascularity are normal. Detectable difference in the degree of vascularity occurs only when severe unilateral stenosis is associated with increased pulmonary blood flow.

Pulmonary oligaeamia in chest x-ray was seen in four of our patients. Two had associated severe valvular pulmonary stenosis and two had multiple peripheral P.S.

Most common type of stenosis are isolated main pulmonary artery or proximal left or right pulmonary artery stenosis (Type I and II). We had 12 patients with Type I and 13 patients with Type II stenosis. Type III and IV were seen in 4 patients.

Associated lesions are common with peripheral P.S. Two third of our patients had associated cardiac malformation, most frequent being valvular pulmonary stenosis, followed by atrial and ventricular septal defects and patent ductus arteriosus.

Natural history of peripheral P.S. is similar to that of valvular P.S: mild form will not progress and regression of

gradient with growth has been demonstrated in infants with peripheral P.S. In multiple peripheral P.S. of severe degree progression of obstruction can occur and prognosis is similar to that of primary pulmonary hypertension[7].

Mild to moderate isolated unilateral or bilateral peripheral P.S. (Type I or Type II) usually doe not require any therapy. Very proximal severe obstruction can be tackled by surgery or balloon dilatation. Recently balloon angioplastry and balloon expandable stents were used in peripheral P.S [5,6]. Reports showed satisfactory reduction in gradient following use of balloon with 3 to 4 times the diameter of the stenosed segment and using prolonged dilatation at 5 to 7 atm of pressure.

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Table I

TABLE SHOWING DISTRIBUTION OF CASES ACCORDING TO GAY'S CLASSIFICATION

Peripheral PS Type	Number
I	12
II	13
III	2
IV	2

Table II

TABLE SHOWING ASSOCIATED ANOMALIES

Associated defect	Number
Pulmonary valvar stenosis	10
Ventricular septal defect	3
Atrial septal defect	3
Patent ductus arteriosus	4
Total	20