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## Surgical Outcome in Total Anomalous Pulmonary Venous Connection Repair in Pediatrics

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Total anomalous pulmonary venous connection (TAPVC) is an uncommon cardiac abnormality with a high potential for mortality unless prompt treatment is provided. We studied clinical characteristics and surgical outcome in children with TAPVC in Rajaei hospital. This study was performed in 41 patients. 30 were male and 11 were female, age ranged from 1 month to 168 months (mean 35.6 months). Abnormal anatomic connection was supra cardiac in 20(46.8%), cardiac in 18(43.9%), infra cardiac in 2 (4.9%) and mix in 1(1.4%). Surgical correction was performed on 30 patients. 8 of the patients died before surgical correction. Early postoperative mortality was in 10(33.3%). No significant relation existed between mortality and sex, type of TAPVC, respiratory infection and congestive heart failure in the deceased patients. Among the survivors (mean follow up 31.5 months), 14 are well without any residual, 1 with permanent pacemaker, 2 with Pulmonary vein obstruction and the remainder with no significant residual. Although surgical correction of TAPVC is a treatment of choice and many centers now achieve reproducibly good results with all anatomic subtypes, in developing countries it has still been associated with high mortality. Better results can be obtained after aggressive preoperative stabilization and postoperative management of patients.

**Key words:** Total anomalous pulmonary venous connection , surgical correction, outcome

## INTRODUCTION

Congenital heart disease (CHD) is one of the many factors that threaten the health of children. The prevalence of CHD is 5.8 in 1000 live births. Total anomalous pulmonary venous connection (TAPVC) is a rare CHD, which in that the pulmonary veins drain to a site other than the left atrium. This anomaly comprises about 1.5% of CHD. TAPVC commonly has an early presentation and requires an operation during neonatal period<sup>[1,2]</sup>. Without surgery 80% of the patients die during infancy. TAPVC usually is an isolated disease but rarely transposition of great arteries and tetralogy of Fallot may be associated disease<sup>[3]</sup>. As studied about this disorder are limited in Iran, therefore the present investigation was performed to evaluate the results of the surgical correction of TAPVC in heart center of Rajaei in Iran.

## MATERIALS AND METHODS

This is a descriptive study, which includes 41 patients admitted in this center with TAPVC. The patients had been admitted from 1990 to 2000 in the pediatric ward and had been followed in the outpatient clinic up to the time of the study. Data had been gathered with reference to the patients, files and the completion of questionnaires. Results have been analyzed in a descriptive manner and the median, mean and frequency have been calculated and presented with tables. Also related factors to mortality analyzed with qui-squire test and  $p < 0.05$  has been considered as significant.

## RESULTS AND DISCUSSION

Forty one patients had inclusion criteria for the study. 30 (73.2%) patients were male and 10 (26.8%) patients were female. Age at presentation ranged from 1-720 days (mean 100 days), age of patients at admission ranged from 6 days to 168 months (mean 36.8 mon) and age of patients at operation ranged from 1-168 months (mean 35.6 mon). Type and frequency of signs and symptoms showed in Table 1. The prevalence of TAPVC at this center in patients who requires surgery with CHD was 0.4%. The type and frequency of all patients with TAPVC has been showed in Table 2.

In electrocardiography, the patients had right ventricular hypertrophy and right atrial enlargement. In chest radiography, the most prominent sign was cardiomegaly and increased pulmonary vascular markings. 20% of patients with supracardiac type of TAPVC had the "snowman" appearance or "figure 8" at chest roentgenography. Definite diagnosis was performed

Table1: Types and frequencies of signs and symptoms in TAPVC

Signs & symptoms	Frequency	Relative frequency (%)
Heart murmur	36	87.8
Tachypnea	32	78.0
Cyanosis	29	70.7
Low weight	22	53.7
Digital clubbing	10	24.4
Respiratory infection	5	12.2

Table 2: Types and frequencies of TAPVC

Type	Frequency	Relative frequency (%)
Supracardiac	20	48.7
Cardiac	18	43.9
Infracardiac	2	4.9
Mix	1	2.4
Total	41	100.0

Table 3: Outcome in operated & unoperated patients of TAPVC

Patients condition	Alive	Dead	Total
Operated	20	10	30
Unoperated	2	9	11
Total	22	19	41

with echocardiography and angiography in 98% of patients before surgery. The most common associated anomaly was atrial septal defect (ASD). The other associated anomalies as decreasing order of frequency were patent ductus arteriosus (6 cases), pulmonary valve stenosis (2 cases), peripheral pulmonary stenosis (1 case), cortriatriatum (1 case), ventricular septal defect (1 case) and coarctation of aorta (1 case). 8 patients died before surgery and 3 patients refuse from surgery. Of the 30 patients who had undergone surgical correction, 10 patients died in early days after surgery (Table 3). In remainder 4 had transient arrhythmia, 1 had surgical complete heart block who need permanent pace maker and the others had not any significant problem immediately after surgery. The mean period of follow up after surgery was 31.5 mon. At follow up of corrected patients, 1 patient had small residual ASD and 2 patients had stenosis of pulmonary veins 2-8 weeks after surgical correction that need surgical correction and the other patients had not significant residual.

At follow up of uncorrected patients, 1 patient died and 2 patients had severe pulmonary hypertension.

Some mortality related factors like congestive heart failure, pulmonary infection, type of TAPVC, low weight and pulmonary hypertension was analyzed in this study but only low weight and pulmonary hypertension had significant relation to mortality ( $p < 0.019$ ,  $p < 0.007$ , respectively).

TAPVC is a rare anomaly and includes 1.5% of CHD. This malformation has an early presentation and often requires an immediate surgical correction during the neonatal period. Of the 41 patients 30 were male and 11 were female. In a study from Hyde *et al.*<sup>[4]</sup> a male preponderance has also been observed. The age of the

patients was between 1-720 days. In a report from Bando *et al.*<sup>[5]</sup> age distribution was between 12 h to 7.3 years. In this study, most cases were of a supracardiac type, which has also been reported from other studies<sup>[6,7]</sup>.

In electrocardiography, the most prominent data were right atrial enlargement and right ventricular hypertrophy as reported by others<sup>[8,9]</sup>. In chest radiography, most common sign was cardiomegaly with increased pulmonary vascular markings. Also 20% of patients with a supracardiac type had a "snowman" sign in chest x-Ray. This sign has also been observed and reported in other studies. All the patients had the diagnosis made with echocardiography, except one. The diagnosis is made with echocardiogram in 97.5% of patients. The prevalence of pulmonary vein stenosis was 10% at the follow up of patients between 2-8 weeks after surgical correction (mean 5 weeks). 2 patients required reoperation. In a study from Hyde *et al.*<sup>[4]</sup> the prevalence of pulmonary venous stenosis was 11% and the mean time of its occurrence was 40 days<sup>[4,10]</sup>.

The most common congenital anomaly in the operated patients was atrial septal defect as in other studies<sup>[11]</sup>.

Of the 30 patients operated on this study, 10 patients died (33.3%) that almost all of them but one had early deaths in hospital.

It must be stated that mortality has decreased from 30% in early reports to 3-13% in recent studies<sup>[11,12]</sup>. In a study from Bando *et al.*<sup>[5]</sup> the mortality after 1991 was nearly zero.

In our study, the mean follow up period was 31.5 month after surgical correction. Bando *et al.*<sup>[5]</sup> Reports a follow up of 5 years and 4 months.

In many reports some factors reported in relation to mortality like site of abnormal drainage<sup>[13]</sup>, sex<sup>[13]</sup>, age at operation<sup>[14]</sup>, pulmonary vein stenosis<sup>[15]</sup> and pulmonary hypertension<sup>[16]</sup>. In our study among many factors only pulmonary hypertension and low weight had significant relation to mortality ( $p < 0.019$ ,  $p < 0.007$ , respectively).

Atrial arrhythmia in early post operation period observed in 20% of our patients. In other study incidence of early post operation arrhythmia 10-20% has reported by John *et al.*<sup>[17]</sup>, but late post operation arrhythmia also reported by Saxena *et al.*<sup>[18]</sup>. Therefore in long term, electrocardiogram and holter monitoring must be in their follow up protocol.

Finally, it must be stated that TAPVC is a life threatening disease. An early diagnosis and surgical correction is life saving. The mortality of total correction of TAPVC in our country can be compared with the results of total correction in developing countries.

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