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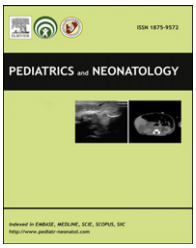

ORIGINAL ARTICLE

Total Anomalous Pulmonary Venous Connection: 15 Years' Experience of a Tertiary Care Center in Taiwan

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Background: Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart disease in which the connection between the pulmonary vein (PV) and left atrium needs to be surgically created. This study investigated the spectrum and outcome of a Taiwanese cohort.
Methods: Isolated TAPVC cases were identified from our institutional database between 1995 and 2009. We reviewed the medical chart and conducted telephone interviews with those lost to follow-up.
Results: There were 78 patients (52% male). The anomalous drainage sites were mainly supracardiac type (42.3%) and cardiac type (39.8%). Before operation, PV stenosis was found in 100% of infracardiac type, and in 69.7% of supracardiac type. Among the 75 patients undergoing operation, the surgical mortality was 9% (7/75). Perioperative arrhythmias (mainly of atrial origin) occurred in 35% of the patients. Of the 68 patients who survived the first operation, 28 (41%) developed pulmonary vein restenosis. Half of them progressed to severe PV stenosis, which required reintervention or resulted in mortality. Preoperative PV stenosis was the most significant predictor for postoperative PV restenosis and PV re-intervention. For the cohort, the 1-year and 5-year survivals were 78.9% and 74.2%, respectively, and the predictor for survival was again preoperative PV stenosis.

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Conclusion: The surgical mortality of isolated TAPVC is now low. Preoperative PV stenosis not only increased the risk of late PV restenosis and its reintervention, but also the overall mortality. The spectrum of PV drainage, *per se*, was not associated with worse outcome. PV restenosis remained the most important issue after correction of TAPVC.

1. Introduction

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart disease, in which pulmonary veins are connected to an abnormal location rather than the left atrium. It accounts for 1% to 3% of congenital heart disease according to Western reports.[1–4] The incidence in Taiwan is 0.11/1000 live births, which accounts for 0.8% of the congenital heart diseases.[5] Since the diagnosis is only rarely made at the fetal stage, the incidence has not changed greatly in recent years. Patients usually develop progressive pulmonary congestion and low cardiac output, and the severity is dependent on the condition of pulmonary venous obstruction (PVO) and the restriction of interatrial communication. Without surgical correction, the prognosis is extremely poor, with a mortality of 80% during infancy.[6,7] With advances in cardiac surgery, the surgical mortality is now in generally below 10%.[1,8] However, pulmonary vein (PV) restenosis may appear after the cardiac repair and may be difficult to manage.[2] This study, based on the 15-year experience of a tertiary medical center in Taiwan, a country with child health index similar to that of the USA,[9] investigated the clinical spectrum of Asian population as well as the predictors of outcomes.

2. Materials and Methods

This study was approved by the institutional review committee of National Taiwan University Hospital, and individual informed consent was waived due to the retrospective nature of the study. Patients with TAPVC were identified from the database of National Taiwan University Hospital from January 1995 to June 2009. The classification of TAPVC was based on that proposed by Darling and associates in 1957 as supracardiac, cardiac, infracardiac, and mixed types.[10] Only cases of isolated TAPVC were enrolled. Except for patent ductus arteriosus, atrial septal defect (ASD), and patent foramen ovale, patients with other associated anomalies (e.g. ventricular septal defect, coarctation of aorta, heterotaxy syndrome, or single ventricular circulation, etc) were excluded from this study. The diagnosis was mainly made by echocardiography. In earlier years, the diagnosis of TAPVC was also confirmed by cardiac catheterization. PVO was defined as peak pulmonary venous flow $\geq$1.5 m/s (roughly equal to pressure gradient 9 mmHg)[11] measured by echocardiography pulse wave Doppler or confirmed by cardiac catheterization, computed tomography (CT; 50% or more narrowing in diameter of PV was considered as PVO), or surgical finding. Of the TAPVC patients, all underwent echocardiography study, 34 were diagnosed only by echocardiography, 22 had additional cardiac CT study, 14 had additional cardiac catheterization study, and 8 had both cardiac CT and cardiac catheterization. Surgical mortality was defined as mortality within 1 month of operation. Patients' medical charts were reviewed, and clinical data collected for statistical analysis. For patients who were lost to follow-up, a telephone interview was performed if possible.

2.1. Statistics

SPSS software was used for statistical analysis. Differences of numeric variables were compared using Fisher's exact and Chi-square tests. Probability of survival after operation was calculated and plotted by Kaplan-Meier method. Logistic and Cox regression analyses were used for multivariate analysis in binary endpoint and time table analyses, respectively. A p-value <0.05 was considered statistically significant.

3. Results

A total of 239 patients were diagnosed with TAPVC. Excluding those associated with other major cardiac anomalies, 78 patients with isolated TAPVC were enrolled into this study. The total follow-up period was 409 patient-years. The basic characteristics of the 78 patients were as follows: the female-to-male ratio was 38 to 41, prematurity accounting for 12.7% (10/79) of the patients. Excluding one patient who underwent cardiac repair at the age of 37 years, the median (mean $\pm$ SD) ages of symptom presentation and diagnosis were 1 day (18 $\pm$ 38) and 9 days (38 $\pm$ 57), respectively. The initial presentations included cyanosis (57%), respiratory distress (43%) and poor activity/appetite (23%). The median age at operation was 24 days (52 $\pm$ 61), with median body weight of 3232 gm (3490 $\pm$ 1104). As shown in Table 1, the most common type of TAPVC was supracardiac (33/78, 42.3 %), followed by cardiac (31/78, 39.8%; among them, 64% had drainage to the coronary sinus, and 32% to the right atrium directly). The mixed type was the least common (4/78, 5.1%); two were combination of supracardiac-cardiac type, and two were mixed supracardiac-infracardiac type. Preoperative PVO was present in 57.7% (45/78) of patients, and most common in the infracardiac type (100%) and least common in the cardiac type (29%).

The flow chart of the clinical events of this cohort is displayed in Figure 1. Of the 78 patients, three did not undergo surgical repair due to family's choice, and all died within one day of diagnosis. For the remaining 75 patients,