Prognosis Stratification Of
Infracardiac Total Anomalous Pulmonary Venous Connection
Base On Perioperative CT Findings

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Introduction
Total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly accounting for 12–5% of congenital heart diseases of which the infracardiac fashion represents approximately in 20% of cases. The infracardiac type is virtually always obstructive, and because of obstruction to pulmonary venous return, these children present in the first few weeks of life with cyanosis, pulmonary edema, and right heart failure. Surgical treatment is almost inevitable but always difficult. Although echocardiography is the first-line imaging modality in diagnosis of infracardiac TAPVC, the diagnosis of this disease entity has remained a tough challenge. Cardiac computed tomography (CT) could help in making a more accurate diagnosis and giving associated details of infracardiac TAPVC. In our institute, the use of CT in assessment of patient with infracardiac TAPVR before and after surgery is substantially increasing in recent ten years. The pre-operative cardiac and vascular anomalies, postoperative anatomical change and the condition of the anastomotic sites could be delineated in a better panoramic view with the developing technology of CT. The purpose of our study was to see possible factors that could be applied to predict their prognosis by using CT.

Material and Methods
A total number of 39 examinations (F : M = 1 : 1.2 : age range 1 day – 11.2 years old; mean 2.3 years old) with echocardiography diagnosis of infracardiac TAPVC out of 4424 cardiac CT examinations during January 1996 to October 2009 were retrospectively reviewed. These were 15 patients (situs solitus n=3; right atrial isomerism n=12), who had undergone surgical intervention and had cardiac CT either for pre-operative assessment, postoperative follow-up, or both. We analyzed their clinical records as well as the image findings to seek any factors that could be used to predict their prognosis. Descriptive statistics was used for analysis.

Result
After reviewing the clinical records and analyzing the images, some possible correlated prognostic factors were brought out as follows: the type of interatrial communication, presence or absence of a small left atrium and left ventricle, ligation of descending vertical vein, pre-operative pulmonary stenosis, and pre- and post-operative pulmonary vein obstruction. Table 1. All 15 patients presented with interatrial communication. According to the image findings obtained on cardiac CT, we further classified the interatrial communication into four types: Patent foramen ovale (PFO), atretic septal defect associated with endocardial cushion defect (ASD with ECD), common atrium without endocardial cushion defect (CA without ECD), common atrium with associated endocardial cushion defect (CA with ECD). Table 2 shows types of interatrial communication and their relations between live and death. Among these prognostic factors, it was pulmonary vein obstruction stenosis and severe case had the worst outcome of the patients. The data suggested the possibility of a worse prognosis if there is still presence of pulmonary vein obstruction (PVO) after surgical intervention (Table 3).

Discussion
Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart disease and classified according to the site where the anomalous connection occurs. In this study, we specified on infracardiac TAPVC. Most of these patients are also combined with other major infraconal anomalies as they often presented with complex heart disease. Many individual factors could affect the outcome of this entity of disease. We proposed and analyzed five possible prognostic factors that maybe useful in clinical practice. Among these factors, there is a significant mortality rate (100%) revealed in patients with post-operative pulmonary vein obstruction (PVO). Meanwhile, the only one patient that underwent ligation of descending vertical vein, came in a result of death with a mortality rate of 100%. This result may not be adopted clinically since there was only one sample. All patients had interatrial communication in different types, and patients having common atrium without endocardial cushion defect had 100% mortality rate. Although there were only about 50% patients out of 15 presented with hypoplastic LA or LV, the mortality rate of this factor is extremely high(86%). Patient with pre-operative pulmonary stenosis also had a high mortality rate(42%) even after surgical correction.

Conclusion
Although total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly that we seldom see in daily practice, we should make efforts to know the pathological anatomy of this disease entity since rapid developing technology of cardiac CT that could delineated in a better panoramic view with multi-planar reconstruction, volume rendering (VR) and maximum intensity projection (MIP). The prognostic factors we mentioned above are highly recommended to apply in our daily practice. Identify these factors on CT imaging in patient with infracardiac TAPVC could assist the clinicians in planning the operative strategy and postoperative care.