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 1.5–3% 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 seek 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 2.0 years old; median 2.7 months old) with proven 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 all undergone surgical intervention and received 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 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(IAC). According to the image findings obtained on cardiac CT, we further classified the interatrial communication into four types: Patent foramen ovale(PFO), atrial 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 that we found significant affected the 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).

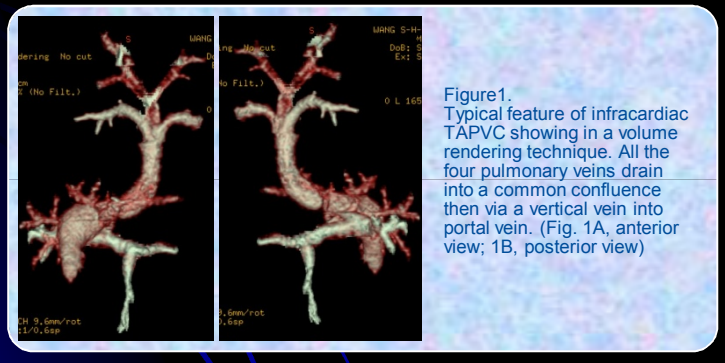

Figure1.
Typical feature of infracardiac TAPVC showing in a volume rendering technique. All the four pulmonary veins drain into a common confluence then via a vertical vein into portal vein. (Fig. 1A, anterior view; 1B, posterior view)

Table1. Infracardiac TAPVC: possible correlated prognostic factors base on perioperative cardiac CT findings

	IAC	LA or LV hypoplasia	PVO	Ligation of DVV	PS
Total No. of patients with the stated prognostic factor(N)	15	7	11	1	12
No. of alive in patients with the stated prognostic factor (until Oct. 2009)	4	1	2*	0	2
No. of mortality in patients with the stated prognostic factor(M)	11	6	9	1	10
Occurrence rate in % (N/T)	100%(15/15)	47%(7/15)	73%(11/15)	6.7%(1/15)	80%(12/15)
Mortality rate in% (M/N)	73%(11/15)	86%(6/7)	82%(9/11)	100%(1/1)	83%(10/12)

T: Total number of patient with Infracardiac type TAPVR, T = 15
IAC: Interatrial communication
PVO: Pulmonary Vein obstruction
DVV: Descending vertical vein
PS: Pulmonary stenosis, including pulmonary atresia
Occurrence rate in % (N/T): The per cent of patient with the stated prognostic factor to total number of patient with Infracardiac type
Mortality rate in% (M/N): The per cent of death with the stated prognostic factor to total number of patients with the stated prognostic factor
*: These 2 patients had pre-operative PVO and successfully corrected after surgery.

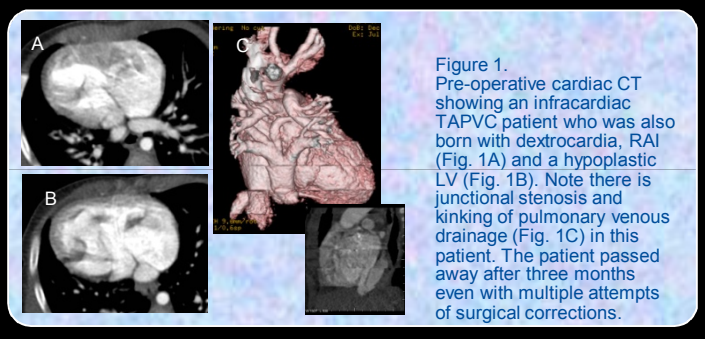

Figure 1.
Pre-operative cardiac CT showing an infracardiac TAPVC patient who was also born with dextrocardia, RAI (Fig. 1A) and a hypoplastic LV (Fig. 1B). Note there is junctional stenosis and kinking of pulmonary venous drainage (Fig. 1C) in this patient. The patient passed away after three months even with multiple attempts of surgical corrections.

Table 2. Type of interatrial communication and their relations between live and death.

IAC type	PFO	ASD with ECD	CA without ECD	CA with ECD
Total number of patient with stated type of IAC(To)	2	1	4	8
No. of alive in patients with the stated type of IAC(until Oct. 2009)	1	0	0	3
No. of mortality in patients with the stated type of IAC(Mo)	1	1	4	5
Occurrence rate in % (To/T)	13%(2/15)	7%(1/15)	27%(4/15)	53%(8/15)
Mortality rate in% (Mo/To)	50%(1/2)	100%(1/1)	100%(4/4)	63%(5/8)

T: Total number of patient with interatrial communication, T = 15
Occurrence rate in % (To/T): The per cent of total number of patient with stated type of IAC to total number of patient with interatrial communication
Mortality rate in% (Mo/To): The per cent of number of mortality in patients with the stated type of IAC to total number of patient with
stated type of IAC

Table 3. Post-operative pulmonary vein obstruction (PVO) and it's relation between live and death.

	Post-operative PVO
Total number of patient with post-operative PVO(Tp)	5
No. of alive in patients with post-operative PVO (until Oct. 2009)	0
No. of mortality in patients with post-operative PVO(Mp)	5
Occurrence rate in % (Tp/T)	45%
Mortality rate in% (Mp/Tp)	100%

T: Total number of patient with preoperative PVO, T = 11(including the 5 cases of post-operative PVO)
Occurrence rate in % (Tp/T): The per cent of total number of patient with with post-operative PVO to total number of patient
with preoperative PVO
Mortality rate in% (Mp/Tp): The per cent of number of mortality in patients with post-operative PVO to total number of patient
with post-operative PVO

Discussion

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart disease and classified according to the site where the anastomosis to the systemic circulation occurs. In this study, we specified on infracardiac TAPVC. Most of these patients are also combined with other major intracardiac 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 was only about 50% patients(7 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(82%) 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've mentioned in this study are 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 post-operative care.