

Survival rate drops substantially in patients with hypoplastic left heart syndrome who possess abnormality in pulmonary vein or left atrium

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Background: Hypoplastic left heart syndrome and its variant (HLHS) are poor prognostic disease group, even now when operative procedures against them have been adjusted better. We predicted that innate properties and pre-operative conditions would make HLHS prognosis worse. We investigated adverse prognostic factors related to HLHS.

Methods: The medical records of 108 HLHS patients (variant 45) were reviewed who were born between 2004 and 2014. All patients underwent Norwood procedure with shunting to pulmonary artery (PA) at a stage prior to Glenn procedure. First, we sought following factors by Kaplan-Meier methods: survival rate, Glenn achievement rate, and Fontan achievement rate. Log rank analysis was performed dividing the whole into two groups by each prognostic factor. Second, we reckoned hazard ratio by Cox proportional-hazards model using prognosis related factors which had significant differences with log rank.

Results: Whole survival rate was 58% in patients who underwent Norwood procedure: Glenn achievement rate was 58%; Fontan achievement rate was 41%. Of 45 dead patients, 82% did not attain to Glenn procedure. Log rank analysis showed that survival rates were decreased by existence of following adverse prognosis-factors; anomalous pulmonary-vein connection or abnormal partition wall in left atrium (abnormal PV/LA: 8% vs. 64%, $p<0.0001$); aortic atresia or mitral atresia (AA/MA: 43% vs. 72%, $p=0.0007$); birth until the year 2010 (48% vs. 79%: $p=0.0082$). Glenn achievement rate was decreased only by abnormal PV/LA (17% vs. 63%: $p<0.0001$). After Cox proportional-hazards model, we obtained hazard ratio of death-rate increase by following factors: 4.9 for abnormal PV/LA ($p<0.001$), 3.2 for AA/MA ($p<0.001$), and 3.2 for birth until 2010 ($p=0.001$). We also obtained hazard ratio of un-accomplishment of Glenn only by abnormal PV/LA (5.5, $p=0.019$). Of 8 patients who died after Glenn procedure, 7 patients possessed abnormal PV/LA, or AA/MA (87% vs. 38%: $p=0.027$).

Discussion: Most of HLHS patients with dead did not achieve Glenn procedure yet. Survival rates were decreased in HLHS patients who owned abnormal PV/LA, or AA/MA. Even after Glenn procedure, patients who owned either of abnormal PV/LA or AA/MA died much more than those without these 2 factors.