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**Outcome of surgical intervention within 24 hours of life for hypoplastic left heart syndrome with intact or highly restrictive atrial septum**

*Ide Y., Ota N., Murata M., Tachi M., Ito H., Kanno K., Ogawa H., Fujita T., Sakamoto K.  
Mt. Fuji Shizuoka Children's Hospital, Shizuoka, Japan*

**Objectives:**The surgical outcome of HLHS has been improving, however HLHS with intact/severe restrictive atrial septum (IAS/RAS) carry a poor prognosis. The purpose of this study is to clarify the surgical outcomes for this cohort.

**Methods:** We performed a retrospective review of 11 patients(8 female) diagnosed as HLHS and its variant with IAS/RAS who required urgent surgery within 24 hours of life from January 2003 to December 2013 at Mt. Fuji Shizuoka Children's Hospital. One patient who combined supra-cardiac TAPVC was excluded from this study. Norwood(NW) procedure was indicated as a first palliation for this cohort before 2007, however ASD creation with bilateral pulmonary artery banding(BPAB) was introduced after 2007 to obtain a quick and secure effect of left atrial decompression with minimal surgical stress on newborn babies. Follow-up data were available for 100% of the patients.

**Results:** Five were diagnosed prenatally and had planed delivery in our institute. Other 6 were born in other hospital and transferred immediately after their birth. Median gestational age and birth-weight was 38(range, 36-41)weeks, 2.41(1.71-3.02)kg, respectively. All 11 patients required urgent operation 7.5(0.75-20)hrs after their birth. Their SpO<sub>2</sub> at OR was 62(43-81)%. As a first palliative surgery, two patients(before 2007) underwent NW procedure and 9 underwent ASD creation with BPAB. After ASD creation with BPAB, prostaglandin-E1 was continued and 4 survived to undergo a NW procedure at a median age of 39(24-54)days. We lost 5 out of 11 in perioperative period of NW procedure due to LOS(2), infection(1), pulmonary emphysema(1), and pneumorrhagia(1). Four accomplished bidirectional Glenn shunt and two of them have already undergone Fontan operation with adequate CVP and SaO<sub>2</sub> during the median follow-up period of 0.59 years(7days-10.2years).

**Conclusions:** HLHS with IAS/RAS is a challenging combination. Our strategy to avoid postpartum NW procedure and to decompress the left atrium as soon as possible after birth by ASD creation with BPAB can bring a better prognosis for this cohort even though prenatally diagnosis is not available.