



SPECKLE TRACKING ECHOCARDIOGRAPHY (STE) IMAGING FOR PREDICTION OF SURVIVAL IN PAEDIATRIC PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION

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Background:

Pulmonary arterial hypertension (PAH) is a progressive disease with varying ages of presentation and is usually irreversible. The primary cause of death relates to deterioration of right ventricular (RV) function. Non-invasive assessment of RV longitudinal systolic strain predicts future right heart failure, clinical deterioration and mortality in patients with PAH. However, its prognostic value for paediatric PAH population is poorly defined. We aimed to use Speckle tracking echocardiography (STE) imaging to assess outcomes in pediatric PAH patients.

Objectives:

To utilize functional ECHO parameters of Right atrium and ventricle to assess outcomes in paediatric patients with Pulmonary Arterial Hypertension.

Methods:

Ours was a prospective observational study done in single centre tertiary care hospital. Patients <18 years with PAH were included in study and those with significant intra/extra cardiac shunt or transient PH were excluded. Study duration:-June 2009-June 2022. STE was used to assess RV function at first visit. Primary end point included all-cause mortality, patients who required Potts shunt or those who were referred for heart and lung transplant. Statistical analyses was done using SPSS 20 software and a p value of <0.05 was considered significant. ROC curve analysis with equal importance to sensitivity and specificity was done and Youden's index was calculated as maximum distance from line of equality.



Results:

Out of total 198 patients in pediatric PAH registry, 155 belonged to Group 1 PAH. 72 patients had significant intra-cardiac and extra-cardiac shunt lesion. Out of the remaining 83 patients, 16 were lost to follow-up or had incomplete data, so a total of 67 patients were included in study cohort. The mean age at diagnosis was 7.56 ± 5.19 years with male:female ratio of 1.09:1. Majority presented with easy fatigue and right heart failure. Median follow-up: -1.16 years (2 months-7 years). Study cohort was broadly divided into 2 groups:- Group 1-Alive(43) and Group 2-Expired(7)/Potts shunt(17) and functional ECHO parameters were compared between the two groups. Mean event free survival of study cohort was 4.2 ± 0.3 years with 1, 3 and 5 year survival of 68%, 64% and 64% respectively. RV Global strain (RVGLS) $< -12.95\%$, RV Free wall strain (RVFWS) $< -16.65\%$, right atrial strain (RA strain) $> 14.25\%$, RV fractional area change (RVFAC) $> 18.7\%$ and RV ejection fraction (RVEF) $> 21.85\%$ were associated with better outcomes. Patients showing better outcome as well as better 1, 3 and 5 year survival had significantly lower RVGLS (p value-0.001) RVFWS (p value-0.023) and significantly higher RA strain (p value-0.002), RVFAC (p value- < 0.001) and RVEF (p value-0.02). RVFWS and RVFAC were found to be most sensitive (82%) and RVEF was found to be most specific (74%) marker for predicting survival based on ROC curve analysis.

Conclusion:

Predicting all-cause mortality and classifying PAH patients aids clinical decision-making. The ability for repeated assessment of RV function enables evaluation of disease progress which is feasible with functional Echo parameters. STE can be a useful marker to predict survival and outcomes in pediatric patients with PAH