

1 **Evaluating the Congenital Heart Surgeons' Society (CHSS) Critical Aortic Stenosis**
2 **Calculator in a New Patient Cohort**

3 Paul J. Devlin MD, Edward J. Hickey MD, Conall T. Morgan MD,
4 Anusha Jegatheeswaran MD PhD, William M. DeCampi MD PhD, William G. Williams MD,
5 James K. Kirklin MD, Eugene H. Blackstone MD, William I. Douglas MD, Luc Mertens MD PhD,
6 Brian W. McCrindle MD MPH
7

8 **Introduction**

9 The CHSS Critical Aortic Stenosis (AS) Calculator predicts the 5-year survival difference
10 between a biventricular (BVR) or univentricular repair (UVR) for a given neonate. It is based on
11 362 neonates enrolled from 1994 – 2001. We evaluated the calculator's accuracy in a new cohort
12 of patients.

13 **Methods**

14 From 2005 – 2013, 246 neonates from the CHSS Critical Left Heart Obstruction cohort were
15 treated for critical AS. Median follow up was 5.8 years, IQR: 2.7 – 7.9. The calculator was used
16 to predict the optimal pathway for each patient. Observed and predicted outcomes were
17 compared by surgical pathway.

18 **Results**

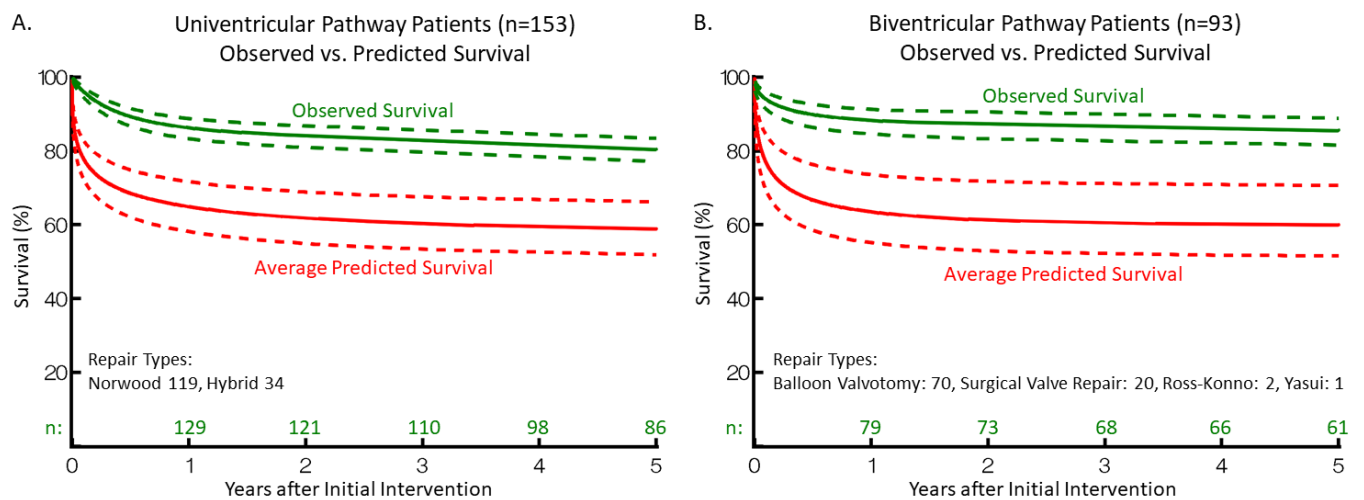
19 Of the 246 neonates, 93 underwent BVR and 153 underwent UVR. Specific repair types in the
20 BVR group were similar to those of the initial cohort, while the UVR group differed due to the
21 use of the hybrid procedure (22%), which was not used in the initial cohort. Five-year survival in
22 the new cohort was 83% (UVR:82%, BVR:86%), improved from the initial cohort (66%,
23 UVR:62% BVR:71%). Comparison of observed patient survival with the model-predicted
24 survival showed that the models underestimate survival in both surgical pathways.
25

26 Twenty-four (16%) UVR patients and 56 (60%) BVR patients were treated discordantly from the
 27 optimal pathway predicted by the calculator. Discordant management and degree of discordance
 28 were not associated with survival. Differences between the new and initial cohorts included more
 29 patients receiving heart transplantation and hybrid operations, a higher number of UVR to BVR
 30 crossovers, higher grades of endocardial fibroelastosis, increased prevalence of mitral stenosis
 31 and left ventricular dysfunction, and lower body surface area.

32 Conclusions

33 The CHSS Critical AS Calculator does not accurately predict optimal surgical pathway in a new
 34 cohort of patients. Over time, survival has improved for neonates with critical AS. Future
 35 patient-specific decision tools should account for changes in patient variables, in management,
 36 and in outcomes.

37 Figure 1



38