

DOWN SYNDROME IN SINGLE VENTRICLE PATIENTS IS ASSOCIATED WITH WORSE SURVIVAL

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Background: We studied the impact of Down syndrome on survival, time to Fontan completion, and survival after Fontan completion in patients with single ventricle physiology.

Materials/Methods: This was a retrospective, multi-institutional study of patients with functional single ventricles who received care at 5 participating centers between January 1992 and December 2014. For the Down syndrome group, we included all patients with Down syndrome and single ventricle physiology. For the control group, we included patients without Down syndrome matched 1:1 to Down syndrome patients by sex, race/ethnicity, and gestational age. Primary outcome was survival. Secondary outcomes included time to Fontan completion and survival after Fontan completion.

Results: A total of 119 patients with a single ventricle were included in the study, with 60 (50.4%) patients in the Down syndrome and 59 (49.6%) patients in the control group. Patients with Down syndrome had similar proportions of systemic right (27/60, 45%) and left (27/60, 45%) ventricles, while systemic right ventricle was more common in the control group (45/59, 76%). At median follow-up time of 7 years, there was 61% survival in the Down syndrome group and 79% survival in the control group ($p=0.007$, Figure, top). On multivariate analysis, Down syndrome was associated with increased mortality ($HR=2.8$, $p=0.005$), while additional cardiac defects ($HR=1.05$, $p=0.4$) and side of systemic ventricle ($HR=0.73$, $p=0.4$) were not. At 7 years, there was 37% cumulative incidence of Fontan completion in the Down syndrome group and 74% in the control group ($p=0.004$). At 5 years after Fontan completion, there was 79% survival in 15 Down syndrome patients who underwent Fontan completion and 97% survival in 38 control patients who underwent Fontan completion ($p=0.03$, Figure, bottom).

Conclusions: Down syndrome was associated with worse survival, lower rates of Fontan completion, and worse survival after Fontan completion in patients with a single ventricle.

Images / Graph / Table

