

Impact of era, type of isomerism, and ventricular morphology on survival in heterotaxy: implications for therapeutic management

Background

Heterotaxy has been demonstrated to reduce survival. There are several different subgroups of patients, however, and no single study has had a large number of patients and analyzed survival across the different subgroups such as patients born in different eras, patient with right and left isomerism, and patients with biventricular or functionally univentricular hearts. This study pools previously reported data from Kaplan-Meier curves and performs such subgroup analysis.

Methods

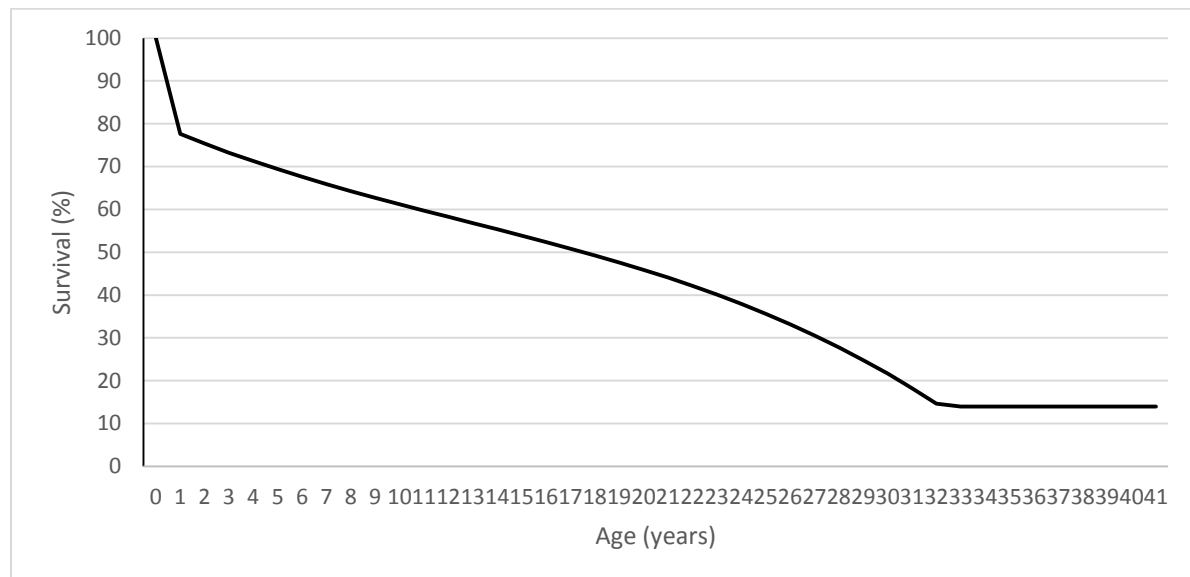
A systematic review of the literature was performed to identify studies reporting survival of patients with so-called “heterotaxy” by means of Kaplan-Meier survival curves. Data was extracted from these survival curves and then pooled together. A polynomial regression was then used to generate a pooled survival curve. This was done for all patients, those born in a more recent era, those with right and left isomerism, and those with biventricular or functionally univentricular hearts.

Results

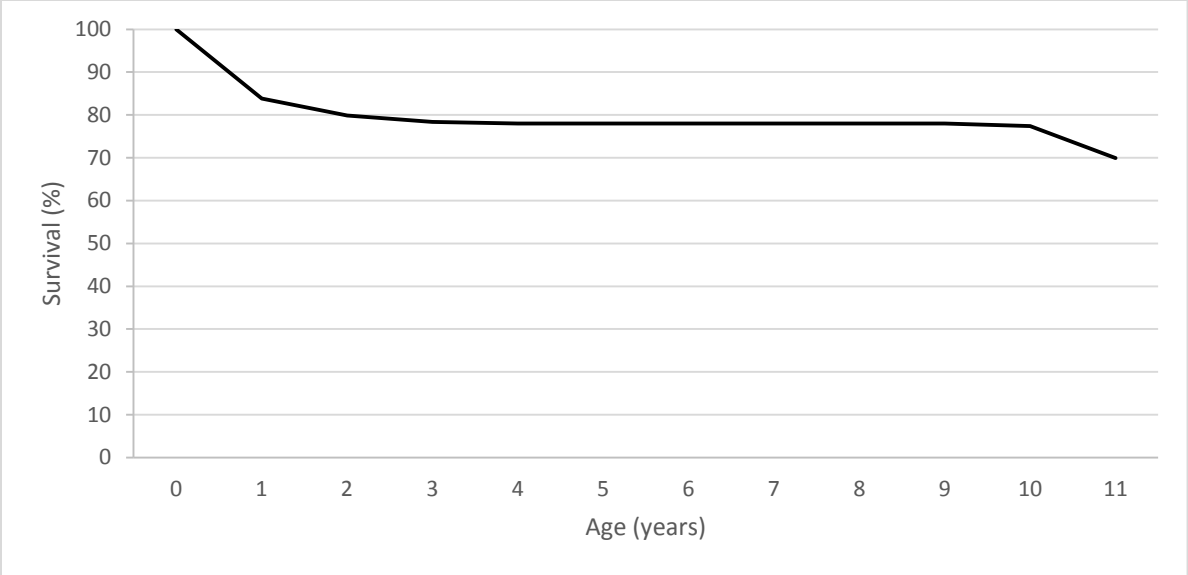
Those born in the more recent era (after 2000) had increased survival compared to the overall cohort. Those with left isomerism tended to have a survival benefit compared to those with right isomerism until about 16 years of age, when those with right isomerism developed a survival benefit. Those with biventricular hearts had a survival benefits compared to those with left isomerism.

Conclusion

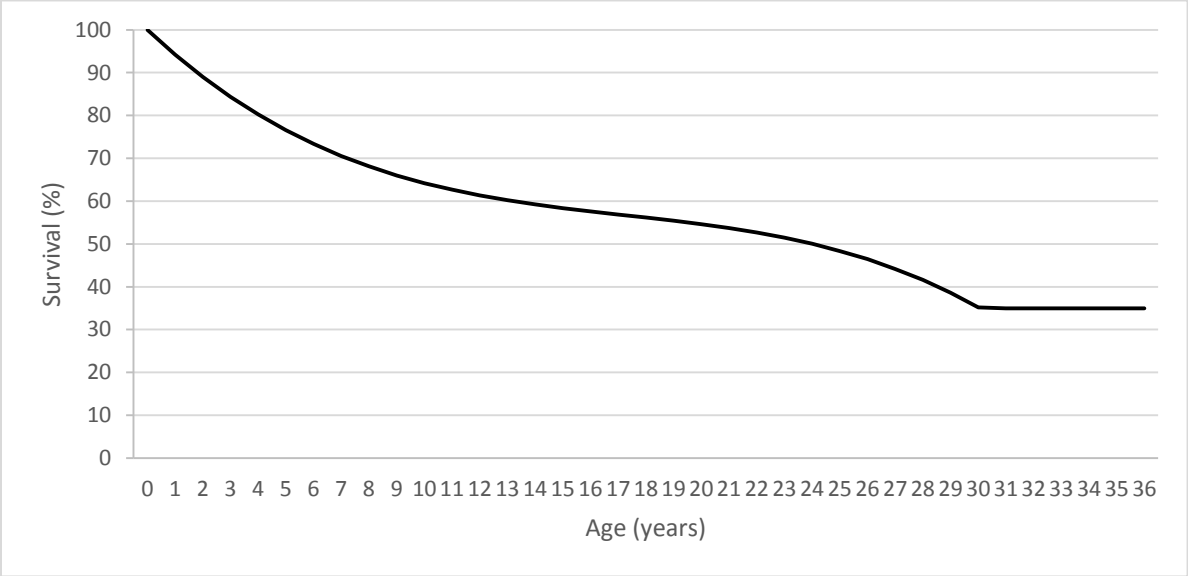
Survival in so-called “heterotaxy” is based on several factors, which include era of birth, sidedness of isomerism, and whether the heart is biventricular or functionally univentricular.



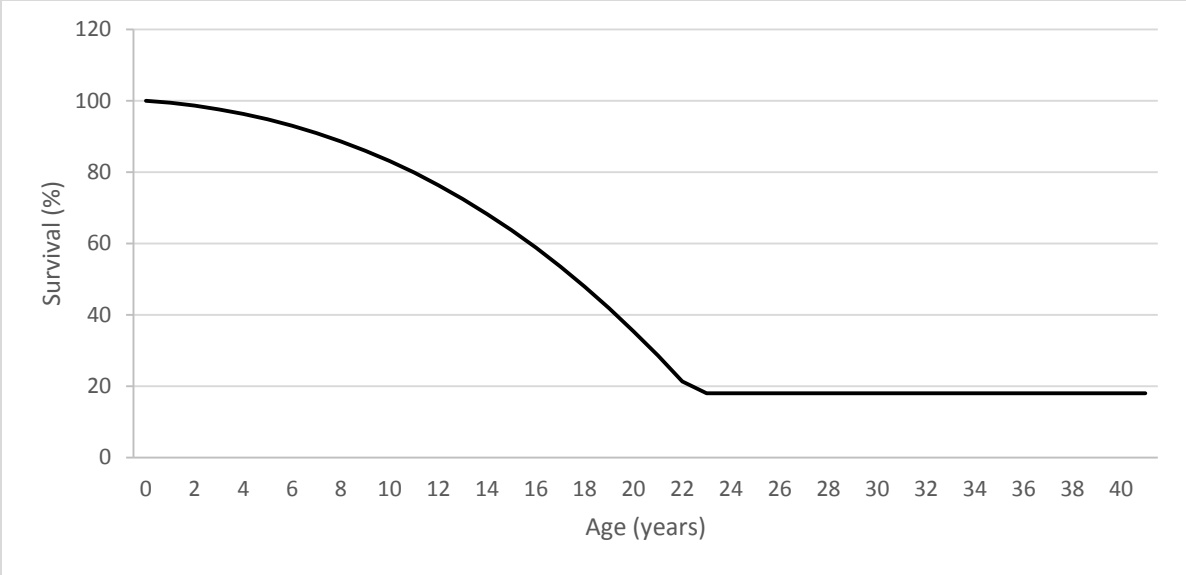
Overall



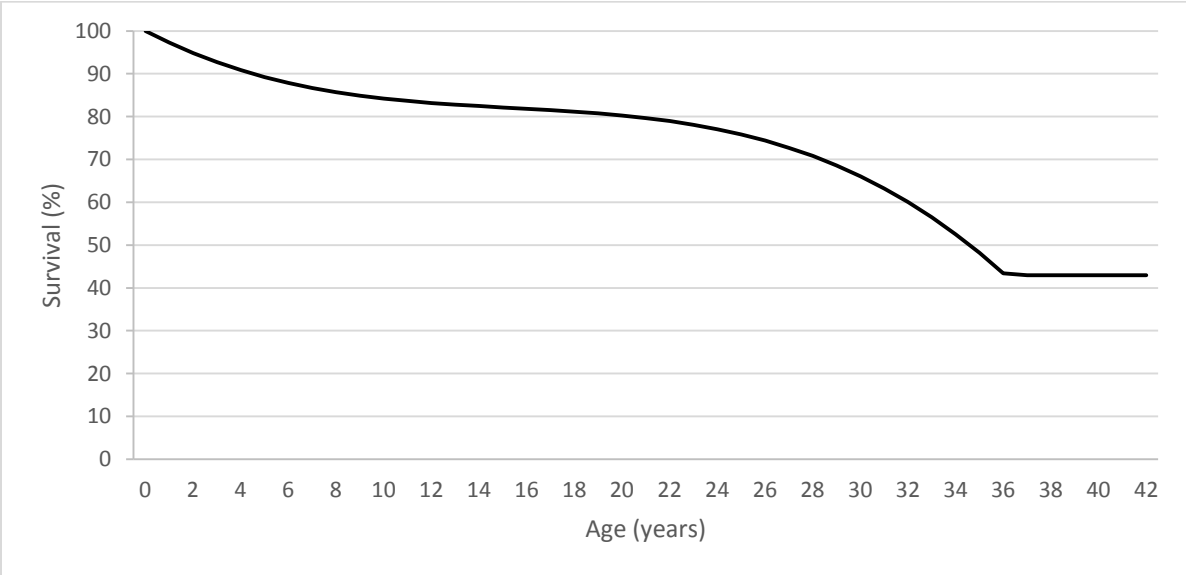
Modern era



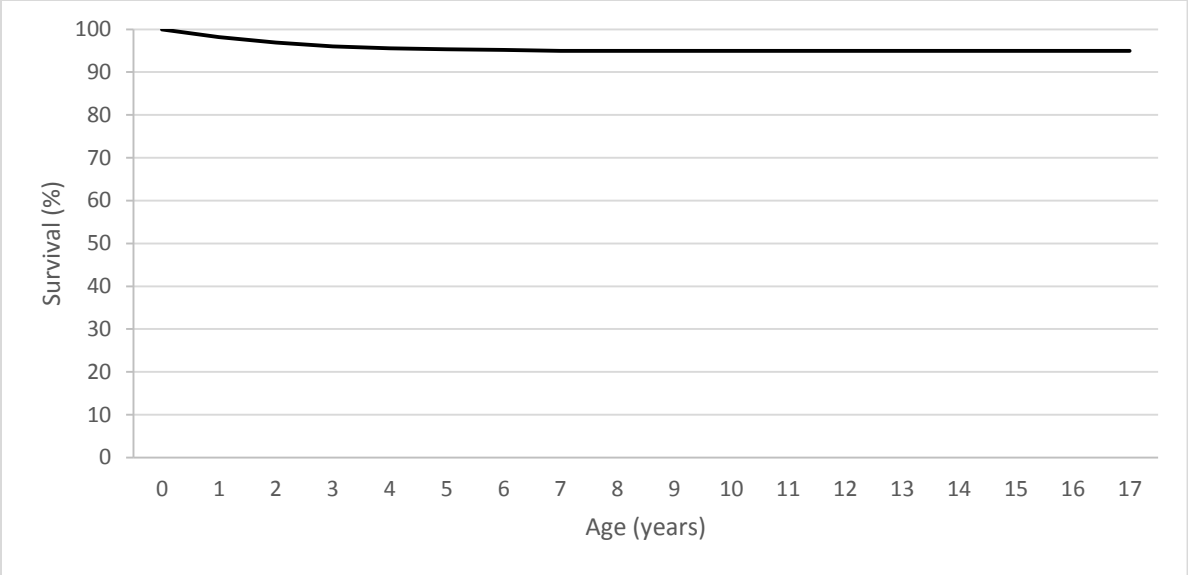
Right isomerism



Left isomerism



Univentricular



Biventricular