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Editorial

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Long-Term Outcomes of Fontan Operation in Korea: Different Regions, Different Patients, Different Prognostic Factors?

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▸ See the article "The Long-term Outcomes and Risk Factors of Complications After Fontan Surgery: From the Korean Fontan Registry (KFR)" in volume 54 on page 653.

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The Fontan operation dramatically extends the lives of babies born with complex congenital heart defects for whom biventricular repair is not feasible. However, due to the physiological limitations of Fontan circulation—such as the absence of a subpulmonic ventricle and elevated central venous pressure—this procedure is associated with numerous long-term morbidities.[1-5] Over the past few decades, significant efforts have been made to address these challenges and improve outcomes.

In this issue of the *Korean Circulation Journal*, Lee et al.[6] report the long-term results of the Fontan operation. This study is one of the largest of its kind, involving more than 1,700 patients from the Korean Fontan Registry over a 40-year period. The majority of patients underwent total cavopulmonary connection, either extracardiac conduit (73.1%) or a lateral tunnel (17.5%). The median follow-up duration was 12.6±8.4 years, with more than 20% of the patients followed for over 20 years. The overall survival rates were 91.7% at 10 years, 87.1% at 20 years, and 74.4% at 30 years. Late mortality and failure risk factors included heterotaxy, significant atrioventricular valve regurgitation, and high pulmonary arterial pressure prior to the Fontan operation.

This study clearly demonstrates a substantial improvement in the outcomes of the Fontan operation over time. The Fontan procedure can now be performed with lower risks. In this paper, the early mortality rate for the extracardiac conduit Fontan operation was 1.3%, gradually improving and lower than that of the lateral tunnel (5.0%) and atrioventricular connection Fontan (14.6%) procedures used in earlier eras. Although the atrioventricular connection type Fontan operation has been identified as a risk factor for early mortality, the improved early surgical outcomes are not solely attributed to surgical modifications. Rather, they are more closely related to the advanced understanding of Fontan physiology and better patient preparation, leading to improved candidacy for the Fontan operation. The long-term outcomes have also shown improvement in more recent eras, with patients who underwent the Fontan operation after 2000 having a higher probability of both short- and long-term survival. However, as other research has shown, non-negligible incidences of long-term morbidities were observed: arrhythmia (12.5%), protein-losing enteropathy (4.1%), thromboembolism (3.5%), pulmonary arteriovenous fistula (2.4%), bleeding (2.0%), and renal failure (2.0%). These complications continue to have a negative impact on long-term survival.

Data Sharing Statement

The data required to reproduce these findings cannot be shared as this is an editorial.

The contents of the report are the author's own views and do not necessarily reflect the views of the *Korean Circulation Journal*.

What we should note about this work is that it was national data from Korea. Most studies on the long-term outcomes of the Fontan operation have been published in Western countries.[1-5] This paper is likely the only national data study published from an Asian country to date. It might provide valuable insights into the following questions: Do different regions have different patients, different surgical outcomes, and different prognostic factors?

Heterotaxy syndrome is more prevalent in Asian populations compared to those in North America and Europe. In this Korean cohort, the prevalence of heterotaxy syndrome is 22.8%, while in Japanese multicenter data, it is 21.8%.[7] Meanwhile, its prevalence is 7.7% in the Pediatric Heart Network database[2] and 7% in the Australia-New Zealand registry.[8] There were also notable differences in anatomic diagnoses. Interestingly, in this Korean cohort, unbalanced atrioventricular canal defect was the most common diagnosis, accounting for 20% of the cohort, whereas atrioventricular canal defect or common atrioventricular valve made up only 4–10% of the Western cohorts.[2][3][5][8] This study reveals that the characteristics of Asian Fontan candidates differ from those in Western countries.

Several studies reported that heterotaxy syndrome is a risk factor for poor long-term outcomes of Fontan operation.[1][4] Does the higher prevalence of heterotaxy syndrome negatively impact the outcomes of Fontan operations? The long-term survival rates in this study are comparable to, or even better than, those reported in previous studies.[4][5] However, since many other factors influence the outcomes of the Fontan operation, we cannot definitively conclude that heterotaxy syndrome does not negatively affect the results. While the outcomes were favorable overall, the risk factor analysis identified heterotaxy syndrome as a significant risk factor for Fontan failure and late mortality. Then, does the varying prevalence of heterotaxy syndrome influence the prognostic significance of heterotaxy syndrome? There have been conflicting reports regarding the impact of heterotaxy on Fontan outcomes. Recent large-scale studies have indicated that heterotaxy is not a risk factor. Marathe et al.[8] analyzed 1,540 patients from the Australia-New Zealand Fontan Registry and found that once the Fontan procedure is successfully completed, heterotaxy is not an important risk factor for adverse long-term outcomes. Downing et al.[5] reported similar findings from Philadelphia. In this large Asian cohort, heterotaxy syndrome was a strong patient-related prognostic factor. Different patient populations may yield different risk factor analyses.

This work demonstrates that the outcomes of the Fontan operation have improved significantly over the past few decades, though there remains room for further enhancement. It also highlights that patients in Asia exhibit different characteristics compared to those in Western countries. A comparative analysis of reports from various regions will help refine the prognostic factors for Fontan operation, providing a foundation for further improving outcomes. We look forward to reports from other parts of the world.

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