Study finds survival rates have improved among heart patients with Down syndrome

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Since 1990, the mortality rate for children born with Down syndrome and a congenital heart defect has more than halved. This has been shown in a study by the University of Gothenburg. However, the mortality rate

in this group is still 85% higher, compared to others who have a congenital heart defect but were not born with Down syndrome.

Congenital heart defects are very common among people with Down syndrome: almost half of those born with the syndrome have a congenital heart defect. The most common is an atrioventricular septal defect, which simply means that there is a hole between the heart's atrium and ventricles.

Turning point in 1990

The research leader behind the study is Zacharias Mandalenakis, who is an Associate Professor at the Sahlgrenska Academy at the University of Gothenburg, and Senior Consultant in Cardiology at Sahlgrenska University Hospital.

"In the 1970s and 1980s, operations were not often performed on heart patients with Down syndrome, and there were probably several reasons for this: these children were considered to have a generally poor prognosis, surgical techniques were still under development, and the heart-lung machine had not been adapted for newborns," explains Zacharias Mandalenakis.

"These conditions received worldwide attention in around 1990, at a time when cardiac care was undergoing rapid development. Heart patients with Down syndrome were then given practically the same priority as everyone else. We can see that this has had a very positive effect on the survival rates for this patient group, which is great."

Mortality rate remains higher

The study has been published in the *Journal of the American Heart*

Association, and is based on registry data from the whole of Sweden for people with Down syndrome and congenital heart defects born between 1970 and 2017.

A total of 3,285 patients with Down syndrome and congenital heart defects, and 64,529 patients with heart defects but without Down syndrome, were included. Each of those with Down syndrome was also compared with eight matched controls—i.e., people from the general population of the same age and gender, but without Down syndrome or congenital heart disease.

The study shows that people with Down syndrome and heart defects have a much better survival rate, although the mortality rate for this group remains increased. Children born before 1990 with Down syndrome and heart defects had a mortality rate that was about 47 times higher than children born without either Down syndrome or a heart defect.

For children born since 1990, the mortality rate has more than halved. This is now 18 times higher for people born with Down syndrome and heart defects, compared to the normal population.

Evaluating the follow-up

For the entire period 1970–2017, people with Down syndrome and congenital heart disease had a 25-fold greater mortality compared to the control group (who had neither Down syndrome nor heart disease). Compared to other people with heart defects, the mortality rate in the Down syndrome group was doubled throughout the period. In the latter part of the period, after the turning point in 1990, the mortality rate among those with Down syndrome was 85% higher, compared to others with a congenital heart defect.

"The care for patients with Down syndrome and congenital heart defects has improved, but it needs to be even better," concludes Zacharias Mandalenakis.

"The Down syndrome patient group can now live longer than before, which may mean that they have time to develop other acquired diseases, in addition to any heart defects. More studies are needed to clarify the reasons for the remaining increased mortality among this group of patients. We also need to evaluate whether these patients may need better follow-up."

More information: Stella Engsner et al, Impact of Down Syndrome on Survival Among Patients With Congenital Heart Disease, *Journal of the American Heart Association* (2024). DOI: 10.1161/JAHA.123.031392

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