Good survival of 'blue babies' and children with congenital heart defects

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Over 90 percent of those operated on for congenital heart defects as children, for example, due to blue baby syndrome, are alive 20 years post-surgery. A new doctoral thesis at Sahlgrenska Academy has explored this issue.

Many congenital heart defects are caused by the connection between the right chamber and the pulmonary circulation being underdeveloped, or even non-existent. The most common group with these heart defects are sometimes called "blue babies" as the heart defect gives the child's skin and mucous membranes a bluish tinge.

Life-long checkups

The problem is treated by surgically creating a new connection, a so-called conduit. The conduit's longevity is, however, limited, which constantly requires new operations and life-long patient checkups.

Researchers at Sahlgrenska Academy have now studied the survival and quality of life of 1 000 patients with congenital heart defects, of which a total of 574 persons had a conduit.

"Our first insight is that the number of adult patients with a conduit has quadrupled during the 2000s. The increase is due to more patients being transferred from pediatric healthcare as well as a dramatic increase in the number of conduit operations over the period," says PhD student Kristofer Skoglund who will be presenting these findings in his thesis.

Low mortality rate

Studies show that the long-term survival of "blue babies" and other patients with congenital heart defects is reasonably good. Over 90 percent of the patients are alive 20 years after the first conduit operation, while the mortality rate within 30 days after the operation is less than 1 percent, reoperations included.

"The majority of the patients that died during the course of the study, died of heart disease. Increased age at first surgery is a factor that is associated with increased mortality," says Kristofer Skoglund.

Self-related health

Another insight from the study is that the patients self-rated health deteriorates over time. Quality of life, however, does not deteriorate

with repeated procedures.

"Even if long-term survival is relatively good, it is limited by heart-related death. The need for new procedures to ensure good heart function is, therefore, great. At the same time, our studies show that the conduit's longevity is shorter after reoperation than after the initial operation," says Kristofer Skoglund,

"It is troubling that the number of patients that need and undergo repeated procedures continuously increases. However, the study shows that nearly half of the patients can be offered reoperation with catheter borne technology.

Close collaborations necessary

The actual study is one of the world's largest on the subject and is one of only a few that focuses on the adult patient. In order for this growing group to be best taken care of, the thesis notes that a close collaboration between pediatric and adult healthcare is necessary.

The thesis, Reconstruction of the RVOT with a conduit: lifetime followup, was defended at a disputation on February 5.

More information: Link to the thesis: http://hdl.handle.net/2077/40889

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