

Left-sided obstructive cardiac lesions in the fetus and the neonate

Akademisk avhandling som för avläggande av medicine doktorsexamen vid Sahlgrenska akademien, Göteborgs Universitet kommer att offentligens försvaras i föreläsningssal Tallen, Drottning Silvias barn- och ungdomssjukhus, Göteborg,

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Avhandlingen baseras på följande delarbeten:

- I. Öhman A, Strömvall-Larsson E, Nilsson B, Mellander M. **Pulse oximetry home monitoring in infants with single ventricle physiology and a surgical shunt as the only source of pulmonary blood flow.** *Cardiology in the Young.* 2013;23:75–81.
- II. Gardiner H, Kovacevic A, Tulzer G, Sarkola T, Herberg U, Dangel J, Öhman A, Bartrons J, Carvalho J, Jicinska H, Fesslova V, Averiss I, Mellander M and Fetal Working Group of the AEPC. **Natural history of 107 cases of fetal aortic stenosis from a European multicenter retrospective study.** *Ultrasound in Obstetrics and Gynecology.* 2016; 48:373-381.
- III. Kovacevic A, Öhman A, Tulzer G, Herberg U, Dangel J, Carvalho JS, Fesslova V, Jicinska H, Sarkola T, Pedroza C, Averiss I, Mellander M, Gardiner HM. **Fetal hemodynamic response to aortic valvuloplasty and postnatal outcome: a European multicenter study.** *Ultrasound in Obstetrics and Gynecology.* 2017. doi: 10.1002/uog.18913
- IV. Öhman A, El-Segaier M, Bergman B, Hanseus K, Malm T, Nilsson B, Pivodic A, Rydberg A, Sonesson SE, Mellander M. **The changing epidemiology of hypoplastic left heart syndrome. Results of a national Swedish cohort study.** (Submitted).
- V. Öhman A, El-Segaier M, Bergman B, Hanseus K, Malm T, Nilsson B, Pivodic A, Rydberg A, Sonesson S, Mellander M. **Transplantation-free survival and risk factors for death or heart transplantation after Norwood surgery in a complete national cohort of patients with HLHS in Sweden 1993–2010.** (Submitted).

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Abstract

Introduction Hypoplastic left heart syndrome (HLHS) is a severe cardiac malformation, fatal in the neonatal period in the absence of immediate care. Palliative surgery for HLHS has been available in Sweden since 1993. The outcome has improved over time, but there is still significant mortality. It has been suggested that fetal valvuloplasty in fetal aortic stenosis may prevent progression to HLHS. Home monitoring of oxygen saturation has been suggested as a method to improve survival after the initial surgery.

Aims The aims were to investigate the survival rate of patients born with HLHS in Sweden from 1990 to 2010, and to evaluate fetal valvuloplasty of the aortic valve as a method of preventing HLHS. A third aim was to evaluate the importance of home monitoring as a method to improve survival after the initial surgery.

Methods The complete national cohort of patients with HLHS was identified through national databases. Changes in incidence and transplantation-free survival were calculated and analyzed in relation to risk factors for death. The natural history of fetal aortic stenosis and the efficacy of a fetal intervention were investigated in two retrospective multi-center studies. Home monitoring was evaluated in an experimental study and survival was compared with a historical cohort.

Results and conclusions The overall 10-year transplantation-free survival of patients with HLHS increased from 40 % 1993–2000 to 63 % 2001–2010. Female gender and was identified as a risk factor. The incidence at birth decreased from 15.4 to 8.4 per 100,000. The proportion of liveborn neonates with HLHS undergoing surgery increased from 50 % to 70 %. Fetal intervention with balloon dilatation of the aortic valve improved postnatal survival but did not prevent progression to HLHS. Home monitoring of oxygen saturation was considered lifesaving in a number of individuals but there was no statistical difference in survival compared to a historical cohort.

Keywords: Hypoplastic left heart syndrome, aortic valve stenosis, fetal heart, fetal therapies, outcome studies, survival analysis, epidemiology, incidence, prenatal diagnosis, pregnancy outcome.

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