The changing epidemiology and outcome in hypoplastic left heart syndrome in Sweden. Results of a national cohort study 1993-2013

1. Department of Women’s and Children’s Health, Karolinska Institutet, Stockholm, Sweden
2. Children’s Heart Center, Skåne University Hospital, Lund, Sweden
3. Queen Silvia Childrens Hospital, Sahlgrenska University Hospital, Gothenburg, Sweden
4. Department of Clinical Sciences, Pediatrics, Umeå University, Umeå, Sweden

Introduction:
In Sweden pediatric cardiac surgery was centralized in 1993 to Lund and Gothenburg and Norwood surgery for HLHS has been offered in both centers since 1993. Since then postoperative survival and prenatal detection rate has gradually increased.

Objective:
To describe the changing epidemiology and outcome in hypoplastic left heart syndrome (HLHS) on a national level.

Methods:
This is a retrospective national cohort study covering all of Sweden. Registry data were collected on all patients with HLHS undergoing Norwood surgery 1993-2013 and on all live-borns with HLHS for the period 1997-2010. The prevalence at 18 weeks gestation was calculated from postnatal and fetal registry data from the western part of Sweden and used as an estimate on the national level. A diagnosis of HLHS required all of the following: 1/ mitral atresia and/or aortic atresia, 2/ hypoplasia of the ascending aorta, 3/ severe hypoplasia of the left ventricle, 4/ no additional cardiac defects. No cases were lost to follow-up.

Results:
139 patients with HLHS had Norwood surgery 1993-2013. 79/139 (57%) were alive in August 2014. Transplantation-free survival increased for each 5 year period (p=0.0011) and the 5-year postoperative survival probability for patients born 2008-2013 was 88% for boys and 73% for girls. Significant risk factors for death were lower birth weight (p=0.0023) and female sex (p=0.0013).

The prevalence at 18 weeks gestation was estimated at 20/100,000. Based on this there were 280 fetuses with HLHS in Sweden 1997-2010, 141 were born alive and 95 were operated. The prenatal detection rate increased from 37/90 (41%) 1997-2001 to 69/89 (78%) 2007-2010, the proportion who were born alive decreased from 61/90 (68%) to 29/89 (33%) while the proportion of live-borns undergoing surgery increased from 38/61 (62%) to 24/29 (83%). The proportion alive in August 2014 was 15/90 (17%) 1997-2001 and 19/89 (21%) 2007-2010.

Conclusion
The increasing proportion of terminations because of HLHS is counterbalanced by an increasing proportion of live-borns undergoing surgery and by an improving postoperative survival resulting in a largely unchanged number of survivors. These results may have implications for prenatal counselling and for planning of postnatal resource utilization.