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Outcome of Hypoplastic Left and Right Heart Syndrome (HLHS and HRHS) after antenatal diagnosis in South Wales over a seven year period

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Objective: To assess the rate of antenatal detection of HLHS and its outcome both in the antenatal and postnatal period. This review was undertaken to help improve counselling and provide better surgical survival information for expecting parents in the future.

Methods: All cases of Hypoplastic Left and Right Heart Syndrome detected antenatally between January 2002 and December 2008 were included in the study. Fetal medicine and fetal cardiac databases at a tertiary fetal cardiology centre as well as CARIS central database in Wales were utilised to carry out the review. The notes were carefully scrutinised to rule out any confounding variables.

Results: There were 55 cases of HLHS and 15 cases of HRHS in South Wales over this period.

Outcome of antenatal diagnosis:

	Total no.	Antenatally detected	Termination	Still birth	Fetal Loss	Born alive
HLHS	55	50	24	2	0	24
HRHS	15	14	5	1	1	7

Outcome of antenatally diagnosed live births:

	No. of Live births	Died without surgery	Received Surgery	Died following surgery	Alive
HLHS	24	1	23	5	18
HRHS	7	1	6	1	5

Hypoplastic conditions of the heart are often isolated conditions with a low rate of other system abnormalities (12 out of 55 for HLHS and 1 out of 15 for HRHS). The rate of chromosomal abnormality with HLHS is 10% (5 out of 50) and 6.7% (1 out of 15) with HRHS.

Conclusion: HLHS and HRHS are one of the most high-risk lesions in children with congenital heart disease. Antenatal detection rate for both these conditions has been satisfactory at over 90% in Wales. Termination rate remains high despite substantial improvement in survival after surgery. Post surgical survival stands at 78% in our series for HLHS with a maximum follow up of eight years and 83% for HRHS with a maximum follow up of five years. Counselling is essential for parents to make an informed decision.