MP3-1

Long-term fate of children operated for the hypoplastic left heart syndrome in a country with high foetal termination rate and centralized paediatric cardiovascular care

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Objectives: To evaluate long-term results of surgical palliation for the hypoplastic left heart syndrome (HLHS) on a territory (Czech Republic, 10.5 million inhabitants) with a country-wide prenatal detection program (estimated detection rate of HLHS >90 %, termination rate of detected HLHS 87 %) and postnatal care centralized to one paediatric cardiovascular centre.
Methods: Retrospective analysis of all consecutive newborns with HLHS admitted between 1999-2012. Non-obligatory re-interventions were defined as those additional to the 3 stages of the Norwood pathway.
Results: From a total of 65 consecutive newborns with HLHS 13 pts. (20%) did not receive surgical treatment because of parental decision, associated anomalies or non-fulfilment of the indication criteria. 52 patients (prenatal diagnosis in 33 %) were directed to the Norwood pathway (median age/weight 7 days/3.2 kg). Early/total mortality after Norwood stage I was 9.6/19% (10/52 pts.) with a significant risk factor being lower weight at surgery (Cox proportional risk per 1 gram =0.997, CI 0.995-0.990, P<0.001). Between Norwood stage I and II 15 catheter/surgical re-intervention were carried out in 13 pts. (aortic arch narrowing in 10/13). 42 pts. aged median 6.8 months underwent stage II surgery with a total mortality of 4.8% and 18 subsequent re-interventions in 13 patients. Finally, 26 patients aged median 3.9 years underwent Fontan completion with early/total mortality 0 and 8%, resp. The probability of survival at 1/5/10 years of age was 77/77/71 %. Probability of freedom from non-obligatory surgical/catheter reinterventions was 58/45/41 %. At long-term follow-up (median 7.8 years) 37/38 pts. are in NYHA functional class I or II.
Conclusions: Due to high foetal termination rate the population of live-born HLHS patients is biased towards those without a prenatal diagnosis. Despite a highly centralized care, surgical treatment of HLHS is still associated with significant mortality and morbidity. Long-term survivals, however, have an acceptable functional status during childhood corresponding to other groups of patients after surgical palliation for functionally single ventricle.
(Supported by MH CZ – DRO, University Hospital Motol, Prague,Czech Republic 00064203)