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Original research article

Long-term results of children operated for hypoplastic left heart syndrome in Children's Heart Centre

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ABSTRACT

Introduction: Hypoplastic left heart syndrome (HLHS), one of the most serious congenital heart defects, can be surgically paliated using 3 subsequent stages. Long-term results of this approach are reviewed in this study.

Methods: Fifty-two consecutive patients (pts.) operated for HLHS from 1999 to 2012 were evaluated retrospectively.

Results: Norwood stage I operation was performed at the median age of 6.5 days with a total mortality of 19%. Significant risk factor for death was lower weight at surgery (Cox proportional risk per 1 g increase = 0.997, CI 0.995–0.990, $P < 0.001$). Between stages I and II, 15 catheter/surgical reintervention had to be carried out in 13 pts. (aortic arch narrowing in 10/13). Forty-two pts. underwent stage II operation at the median age of 6.8 months with a total mortality of 4.8% and 18 subsequent reinterventions in 13 patients. Finally, 26 patients aged median 3.9 years underwent stage III operation (total cavopulmonary connection) with a total mortality of 8%. The probability of survival at 1/5/10 years of age was 77/77/71%. At long-term follow up (median 7.8 years) 37 of the 38 surviving patients are in NYHA functional class I or II.

Conclusions: Despite a highly centralized care, surgical treatment of HLHS is still associated with a 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.

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Introduction

Hypoplastic left heart syndrome (HLHS) represents one of the most serious complex congenital heart defects (CHD), 100% fatal in its natural course. The only therapeutic option for these children is a staged surgical palliation or heart transplantation. If surgery is contraindicated, or parents refuse surgical treatment, comfort care is offered with inevitable death. Before establishing the nationwide prenatal detection program of CHD in the Czech republic the prevalence of HLHS was 0.21/1000 live births (3.42% of all the CHD). Male gender prevails [1].

Anatomy of the HLHS refers to abnormal development of the left ventricle, aortic and mitral valves in various combinations from atresia to stenosis and aortic arch hypoplasia (Fig. 1). Associated cardiac defects have been reported, such as bicuspid pulmonary valve, cleft of the tricuspid valve, dysplasia of the tricuspid or pulmonary valves, less frequently, total or partial anomalous pulmonary venous return, atresia of the coronary sinus and interruption of the aortic arch [2,3]. Postnatal survival of newborns with HLHS depends on open fetal shunts – foramen ovale and ductus arteriosus.

The first successful surgical treatment of children with HLHS was reported by Norwood et al. in 1980 [4,5]. Since then, development of surgical techniques, cardiopulmonary bypass, and postoperative care improved survival [6,7]. A number of studies have been carried out to analyze the risk factors affecting surgical result [8]. Current surgical strategy comprises of 3 stages. Neonates undergo stage I (Norwood) operation, connecting the right ventricle to both the systemic and pulmonary circulations. Neo-aorta is constructed using the pulmonary trunc and pulmonary flow is established by a vascular prothesis [9,10]. Stage II operation, bi-directional cavopulmonary anastomosis (BCPA), is performed usually by 4 months of age. In this stage, superior vena cava is anastomozed to the right pulmonary artery and the vascular prosthesis is taken down. The procedure reduces volume load to the systemic right ventricle. The final stage III, total cavopulmonary connection (TCPC) is completed by connecting the inferior vena cava to the right pulmonary artery with use of a Gore-tex tunnel. The systemic and pulmonary circulation work in series, with the right ventricle as the only single driving force. This stage is usually performed around the age of 2–4 years and a weight over 10 kg. In patients with lower capacity of the pulmonary vascular bed or elevated end-diastolic pressure in the right ventricle, a fenestration between the systemic venous tunnel and the pulmonary venous atrium is created. An obligatory right-to-left shunt through the fenestration maintains satisfactory cardiac output at the expense of a slight systemic desaturation. The staged palliative correction is only feasible if normal resistance and capacity of the pulmonary vascular bed have been proved.

Due to the progress of the ultrasound diagnostics and teaching programs for gynecologists, prenatal detection of the HLHS at the early stages of pregnancy have exceeded 90% in his country [11]. Number of fetuses diagnosed with HLHS and the number of patients operated at our institution is shown in Fig. 2. At our center, the program of surgical care for patients with HLHS was established in 1999. Retrospective analysis of the results is subject of this paper.

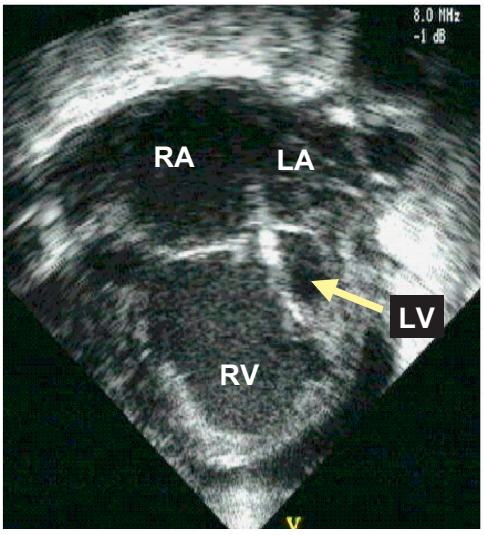

Fig. 1 – Echocardiography of hypoplastic left heart syndrome. Apical four chamber view, (RA – right atrium, LA – left atrium, RV – right ventricle, LV – left ventricle) (with permission of [2]).

Patients and methods

Since the beginning of the program, we employed semi-selective operability criteria (Table 1), to eliminate unfavorable effects of specific anatomic and functional findings on the result of staged palliative surgery [8]. Fifty-two pts. (44 boys, 8 girls) out of a total of 65 newborns admitted with diagnosis of HLHS underwent stage I palliation from January 1999 to the end of 2012. Stage I surgery was performed at the age of 3–17 (median 6.5) days in patients weighting from 2.4 to 4.3 (median 3.2) kg. Delayed sternal closure was used routinely to prevent low cardiac output caused by tissue edema and cardiac pseudo-tamponade. Only 17/52 (33%) newborns were diagnosed prenatally reflecting the fact, that most prenatally

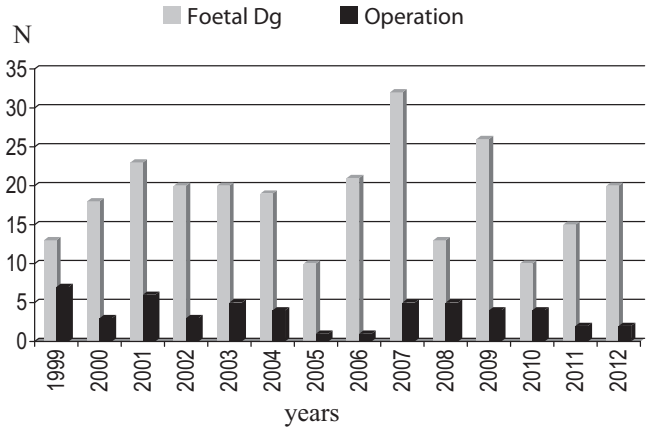

Fig. 2 – Number of fetuses diagnosed with HLHS to number of newborns operated for HLHS.