Hypoplastic left heart syndrome (HLHS) is rare congenital heart defect in which the left side of the heart is severely underdeveloped. It has been a lethal congenital heart anomaly until the last four decades until three palliative surgeries were established (operation by Norwood, Glenn and Fontan). The aim of this study was to evaluate the outcomes of treating patients with HLHS.

The main methods were statistical, and the clinical characteristics were retrospectively reviewed.

We included 132 patients in 20-year period who have been treated at University Hospital Centre Zagreb, operated in our and in foreign centers.

We followed them before, in the meantime and after final operation. Of all patients, 69 survived and 63 died. The highest mortality was in period between operation by Norwood and Glenn in early infancy and accounts for almost 48% all lethal outcomes.

The most common anatomic variant is the mitral atresia and aortic atresia (MA-AA) subtype and the rarest is mitral stenosis and aortic atresia (MS-AA) subtype.

Apart of a three – staged operation procedures, 53 patients required one or more interventions involving implantation of the stent into the pulmonary branches, isthmus the aorta, the Sano or mBT junction, and the dilatation of the same, then coiling of the arteriovenous malformations, electrostimulator implantation and the closure of fenestra. The most common interventions are stent implantation into the pulmonary branches and dilatation of the aortic recoarctation and stenosis of the pulmonary branch stent. In twelve patients, fenestra was closed with an Amplatzer occluder.

The mean follow-up age operation by Fontan (TCPC) is 7.64 years (1,1 – 16,5 years). Two patients were transferred to the GUCH population.

This retrospective study included 132 patients with hypoplastic left heart syndrome in twenty-year period who have been treated at University Hospital Centre Zagreb. The overall survival of all patients is 52.2%. The highest mortality was in period between operation by Norwood and Glenn in early infancy. The most common interventions are stent implantation into the pulmonary branches and dilatation of recoarctation of the aorta. Mean follow-up age operation by Fontan (TCPC) is 7.64 years.

Brain death is frequently associated with systolic dysfunction. The actual mechanisms are not yet completely understood. The goal of this study is to assess echocardiography findings among organ donors in our institution.

Methods we conducted a retrospective study for the period of 17 years (October 2001- December 2018). A total of 20 patients under 18 years with declared brain death were identified. The mean age was 8.8 years and 70% (14 patients) were male. One patient had a history of cardiac disease – ventricular septal defect- and his heart was not accepted for donation.
The adequate transthoracic echocardiogram was possible in 18 (90%) of potential organ donors. All examinations were performed before evaluation protocol confirmed brain death.

Results echocardiogram was normal in 13 (65%) patients. One had a moderate mitral and tricuspid insufficiency. Minimal pericardial effusion was present in 4 (20%) patients. Two had mild septal dyskinesia with normal left ventricular ejection fraction. Diffuse hypokinesis with ejection fraction of less than 55% was found in 5 (37%) patients, in one of which it was less than 45%. A total of 14 hearts (70%) were harvested for transplantation, including the one with the poorest systolic function.

Conclusion mild left ventricular systolic dysfunction occurs frequently in children with brain death, but these hearts can still be considered for transplantation.

Since the affected gene is widely expressed in multiple adult and foetal tissues including gastrointestinal system, brain, lungs, immune system and testis, extracardiac manifestations are common.

The risk for life-threatening ventricular tachycardia is the limiting factor of TS. Since ventricular tachycardia is the leading cause of death in patients with TS, effective anti-arrhythmic medication and an implantable cardioverter defibrillator are the mainstay of therapy. Conclusions Timothy syndrome is a rare congenital arrhythmia disorder with dysfunction in multiple organ systems. Patients are at high risk for sudden death due to life threatening ventricular tachyarrhythmia. Implantation of an ICD at a very young age may be the best means to prevent sudden death.