Hypoplastic Left Heart Syndrome (HLHS)

(Core Condition)

Description	A condition in which the structures on the left side of the heart and the aorta are extremely small, insufficient to support systemic circulation and with normally related great arteries. Classically, this condition includes hypoplasia of the left ventricle, atresia or severe hypoplasia of both the mitral and aortic valves, hypoplasia of the aortic arch, and coarctation of the aorta.
Inclusions	Any diagnosis of hypoplastic left heart syndrome, regardless of whether all conditions in the classical definition are present.
Exclusions	Hypoplasia or diminished size of the left ventricle alone without involvement of other structures on the left side of the heart or the aorta. Hypoplastic left heart or small left ventricle that occurs as part of another complex heart defect, such as atrioventricular septal defect.
ICD-9-CM Codes	746.7
ICD-10-CM Codes	Q23.4
CDC/BPA Codes	746.70
Diagnostic Methods	While hypoplastic left heart may be suspected by clinical presentation, examination, and EKG changes, it may be conclusively diagnosed only through direct visualization of the heart by cardiac echo (echocardiography), catheterization, surgery, or autopsy.
Prenatal Diagnoses Not Confirmed Postnatally	These conditions may be included as cases when only diagnosed prenatally. However, if it is possible to ascertain the degree of certainty of the prenatal diagnosis, this should factor into the decision as to whether or not to include an individual case in the surveillance data, as it may be difficult to distinguish this condition from other abnormalities of the left ventricle prenatally. Live-born children who survive should always have confirmation of the defect postnatally before being included.
Additional Information	NA

