INTRODUCTION. To review literature about long-term outcomes of fetuses with congenital heart disease (CHD).

MATERIALS. Search in PubMed, EMBASE, Medline, Reference lists with no limits of time. Inclusion criteria: follow up of newborns with CHD, data reported as proportional rates. Exclusion criteria: data reported in graphs or percentage, case reports, non-English language reviews, reviews. From each article, age at enrollment, length of follow up, type of CHD, rates of adults died and alive at the end of follow up, cause of death, morbidity were abstracted. Long-term outcomes were defined if they occurred >30 days from birth. CHD were grouped in septal defects, right heart anomalies, left heart anomalies, conotruncal defects, single ventricle and miscellaneous. Cardiac death was divided in heart failure, sudden death, heart surgery, cardiovascular disease, miscellaneous. Extracardiac deaths were not considered for analysis. PRISMA guidelines were followed.

RESULTS. From 10 articles, 17,604 adults were followed up for CHD. The median age at time of enrollment was 26 years and follow up ranged from 5 to 50 years. Out of 17,604 adults, 16,169 (90%) were alive and 1630 (10%) died. Type of CHD was described in 689/1630 (42%) cases. The most frequent was conotruncal defect (26%, 169), followed by septal defects (24%, 169), single ventricle (9%, 66), left heart anomalies (60:9%), right heart anomalies (2%, 13). The remaining 204 (30%) cases were miscellaneous. In 1049 (69%) the cause of death was reported. It was cardiac in 799 (76%) and extracardiac in 242 (23%) cases. In the remaining 8 (1%) cases, the etiology of death remained unknown. Type of cardiac death consisted in heart failure (209, 26%), sudden death (23%, 184), heart surgery complications (14%, 111), cardiovascular complications (13%, 107) and miscellaneous (22%, 174). In 14 (2%) cases, type of cardiac death was not specified. Morbidity was reported in 2 articles and was so heterogeneous that could not be pooled in a systematic review.

CONCLUSIONS. Prenatal counseling of CHD should specify that in fetuses with CHD, long term mortality is 10%. Conotruncal defects seem to be associated with the highest risk of cardiac death in adulthood, which occurs mainly for heart failure. Further studies are needed to investigate long-term morbidity of CHD.