

Prevalence Of Extracardiac Birth Defects And Genetic Conditions In Hypoplastic Left Heart Syndrome And Association With Hospital Mortality

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BACKGROUND

- Up to 30% of patients with congenital heart disease have associated extracardiac birth defects and/or genetic conditions (BD&GC), and the presence of these may influence therapies and outcomes.
- We aimed to evaluate the prevalence of BD&GC in hypoplastic left heart syndrome (HLHS) and evaluate the associations between specific BD&GC and hospital mortality and post-Stage I mortality.

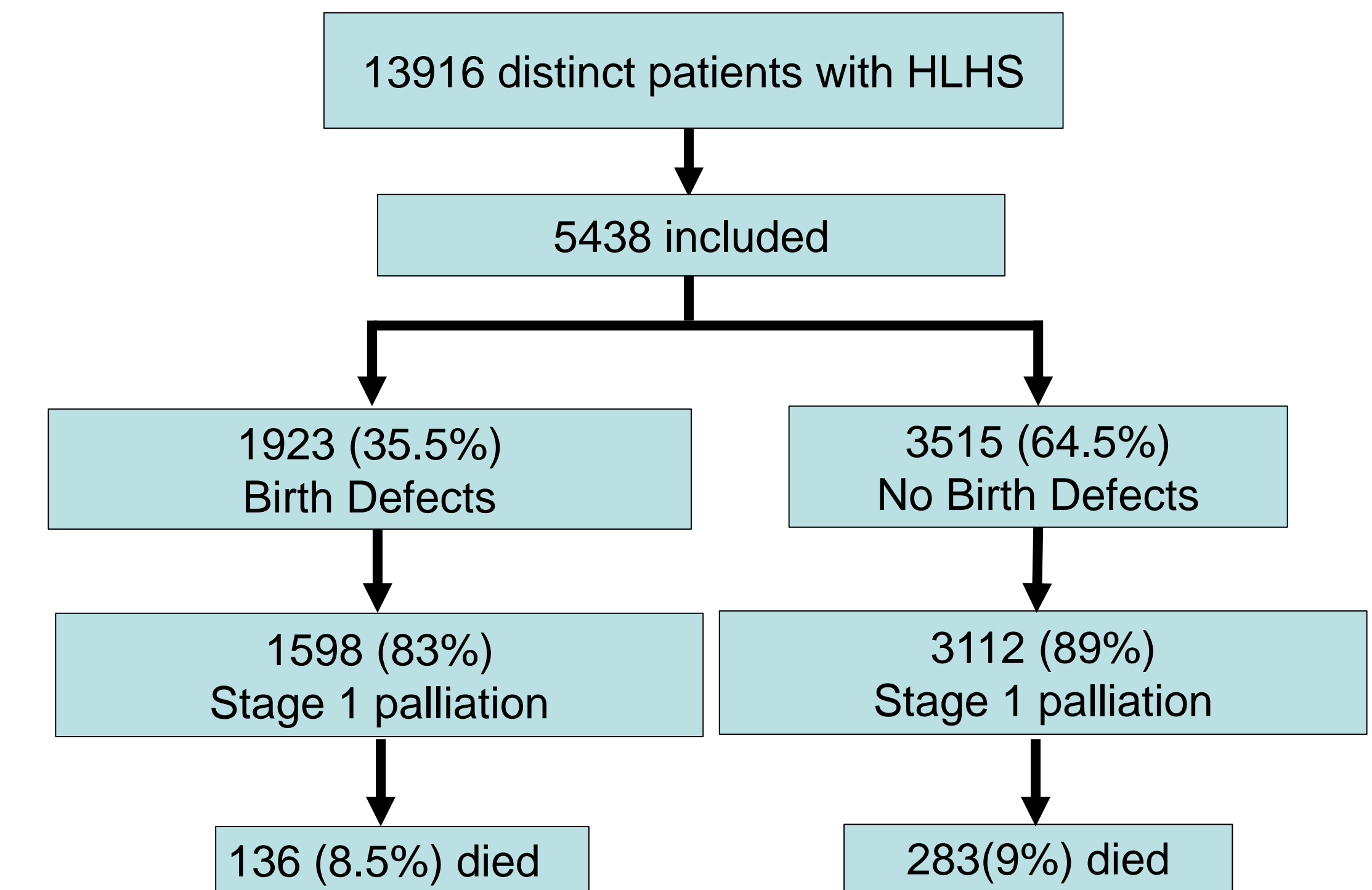
HYPOTHESIS

- Patients with BD&GC have increased mortality.

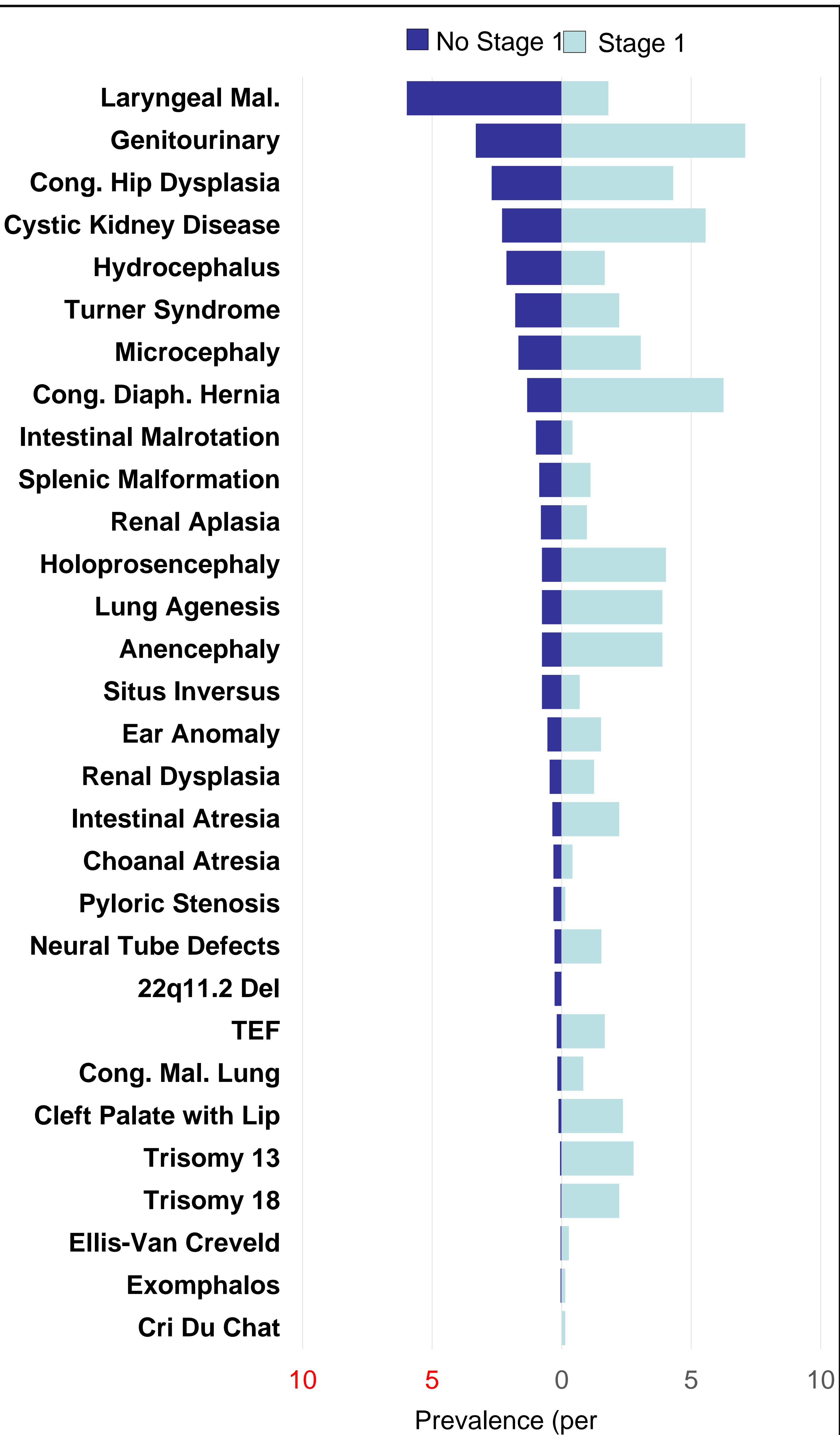
METHODS

- Study design: Retrospective cohort study
- Data: Pediatric Health Information System database from 1999-2018 with an ICD9/10 code for HLHS, no codes for conflicting congenital heart disease, and admission <30 days of life.
- Inclusion criteria:
 - <30 days old and one of the following
 - Stage 1 surgery
 - Hospice
 - Death or transplant
- Statistical Analysis: Pearson Chi square test to analyze the effect of birth defects on mortality.

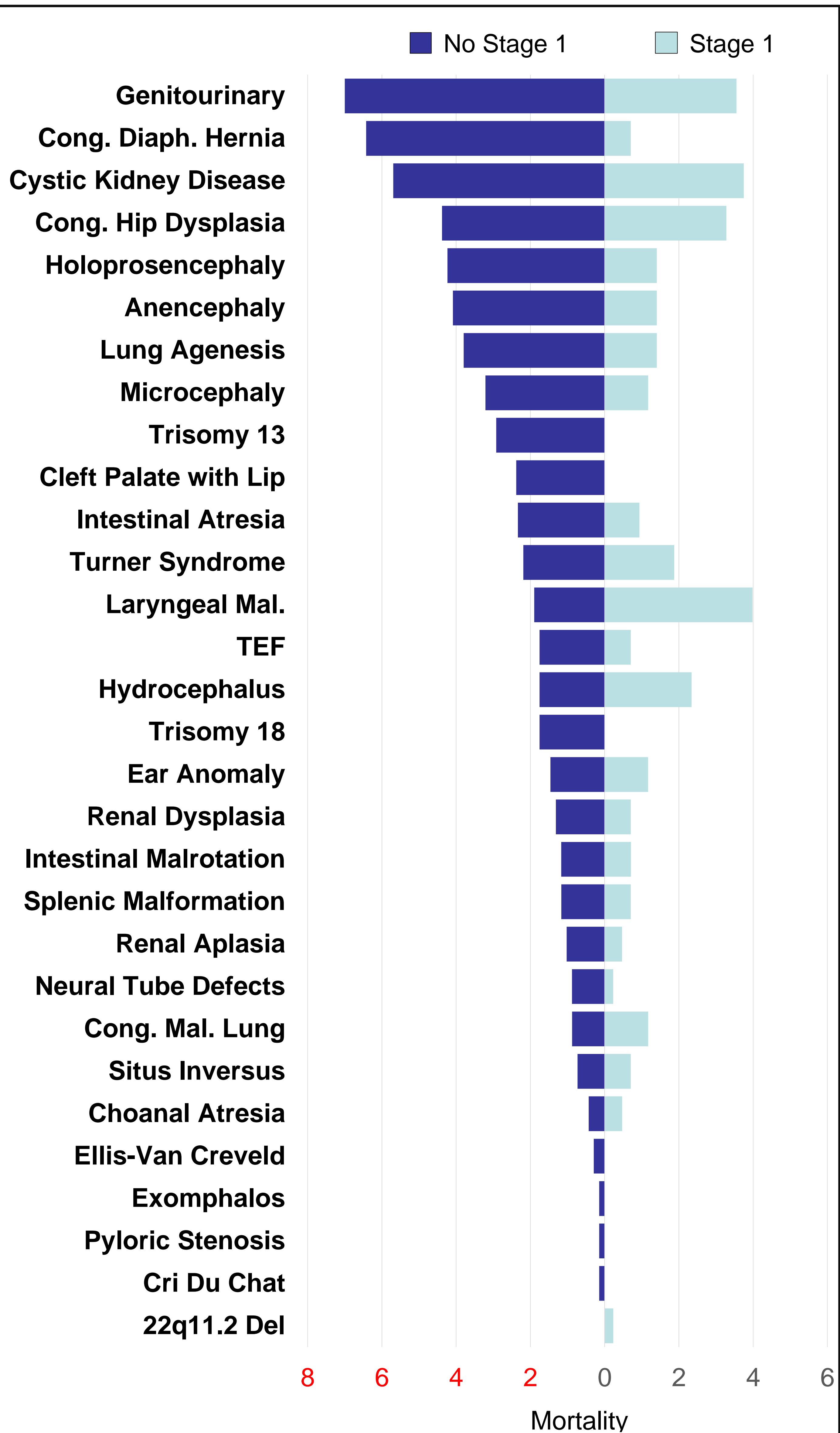
RESULTS



PREVALENCE OF BIRTH DEFECTS & GENETIC CONDITIONS



MORTALITY WITH BIRTH DEFECTS & GENETIC CONDITIONS



LIMITATIONS

- Use of administrative database
- Retrospective study
- Incidence derived from patients admitted to the hospital
- Small number of patients with individual birth defects

CONCLUSIONS

- Most common birth defects in HLHS:
 - Laryngeal anomalies
 - Genitourinary anomalies
 - Congenital hip dysplasia
 - Cystic kidney disease
- Additional mortality due to birth defects varies widely by the condition
- There is no increase in mortality in patients with birth defects who undergo a successful stage 1 palliation
- The associations between HLHS and other birth defects may help elucidate additional genetic etiologies of HLHS

FUTURE DIRECTIONS

- Similar study using Birth Defect Registry Data will better delineate birth defects and genetic conditions associated with HLHS.

REFERENCES

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