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SURGICAL OUTCOMES IN CHILDREN WITH TRISOMY 13

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PURPOSE:
Trisomy 13 is a rare genetic condition with a characteristic set of severe congenital abnormalities. Traditionally, the standard of care was to provide palliative care only, however, there has been a recent shift towards life-prolonging care, including surgery. This study seeks to describe surgical outcomes in patients with trisomy 13 and compare them to comorbidity-matched controls.

METHODS:
Using the ACS NSQIP Pediatric 2012-2019 Participant Use Data Files, patients with trisomy 13 were identified and described. A nearest-neighbor 10:1 propensity score match was performed using demographics, comorbidities, and procedural details. This yielded 254 patients with trisomy 13 and 2,422 controls. Risk ratios for complications and mortality by trisomy 13 status were determined using modified Poisson regression. The primary outcomes were thirty-day mortality and the occurrence of any complication.

RESULTS:
The median age of patients with trisomy 13 was 16 months (IQR 7 years). 126 were male (49.6%) and 128 were female (50.4%). There were no differences in overall complications compared to controls (31.8% vs. 29.7%, RR 1.06, 95%CI 0.87-1.28, p=0.554), but patients with trisomy 13 had markedly higher mortality (7.9% vs. 1.8%, RR 4.43, 95%CI 2.28-8.61, p<0.001). There was no difference in preoperative DNR/DNI status (2.0% vs. 1.7%, p = 0.787). Other complications are listed in Table 1.

CONCLUSIONS:
We conclude that patients with trisomy 13 undergoing surgery have frequent complications and an elevated although not prohibitive risk of death. Compared to patients with similar comorbidities, they have similar rates of complications but a markedly higher risk of mortality. Further research is needed to better understand this discrepancy. Parents of children with trisomy 13 require thorough counseling on these risks prior to making a decision for surgery.