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The outcome of Eisenmenger patients with trisomy 21 does not differ from patients without trisomy 21.

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Introduction: Several patients with trisomy 21 developed the Eisenmenger syndrome (ES) because the underlying congenital heart defect was not corrected. However, little is known about their prognosis. This study aimed at (1) identifying risk factors for worse prognosis in ES patients, and (2) evaluating whether outcome of ES patients with trisomy 21 differs from ES patients without trisomy 21.
Methods: Data on all Eisenmenger patients in follow-up at the pediatric and adult congenital heart disease clinic of the University Hospitals Leuven were collected for retrospective analysis. Regression analysis was performed where applicable and survival rate was compared between patients with and without trisomy 21.
Results: One hundred thirty-four patients (mean age at latest follow-up 33.2±13.6 years, 41.8% male, 44.8% trisomy 21) were included in the study. Complex lesions, right heart failure, impaired renal function, lower transcutaneous saturation and lower body mass index were predictive for impaired outcome. Mean survival of the global ES group was 44.9±2.2 years. However, long-term survival of trisomy 21 patients was not statistically different from patients without trisomy 21 (mean survival 44.5±2.6 years vs 44.5±2.9 years respectively, P=0.80, log rank test).
Conclusions: Long term survival is markedly reduced in Eisenmenger patients. Complex lesions, right heart failure, impaired renal function, lower transcutaneous saturation and lower body mass index were related towards prognosis. However, survival of trisomy 21 patients did not differ from patients without trisomy 21.