Imaging and laboratory markers associated with quality of life in Fontan patients

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Introduction
With the increase in life expectancy of Fontan patients quality of life has become a fundamental aspect of these patients’ follow-up. Recent studies have addressed quality of life in Fontan patients with heterogeneous results. The aim of this study was to assess the importance of variables associated with functional status in quality of life of Fontan patients.

Methods
Fontan patients aged 8 years or older and followed up regularly at our hospital were invited to fill in the Paediatric Quality of Life Inventory 4.0 questionnaire® (PedsQL) at their evaluation at the Fontan Clinics (version according to age). Clinical evaluation, blood tests, trandhoracic echocardiogram (TTE) and cardiopulmonary exercise test (CPET) were also performed. The four main scales for quality of life related to health contemplated in PedsQL - physical, emotional, social and academic functioning were evaluated as well as the summary score for psychosocial health.

Results
Fourty-two Fontan patients aged 9 to 36 years of age (median 18.5 years, IQR 12-23) were included. Sixty per cent of patients were male. Completion of Fontan was performed at a median of 6 years (IQR 5-8), and Fontan completion had been performed at a median of 10 years (IQR 7-15) before this evaluation. Physical functioning was reported as worse for female patients (62.42±15.60 vs 73.69±16.17 points, p=0.0039). Emotional functioning was positively related to global longitudinal strain (GLS) of the dominant ventricle (p=0.026). Social functioning was positively associated with age (p=0.029), with no relation with time since Fontan completion. Academic functioning was positively related to CPET workload (p=0.038). Total quality of life was positively related to age (p=0.032), and negatively related to BNP (p=0.041), with no relation with age since Fontan completion. In a multivariable analysis total quality of life increased 5.14 points for each 10 years of life (p=0.022) and decreased 3.55 points for a BNP increase of 100pg/mL (p=0.028). No variables were related to psychosocial performance.

Conclusions
Variables associated with quality of life in Fontan patients were GLS, BNP and CPET workload. Therapeutic approaches in Fontan patients with an impact on these variables may have a positive influence on quality of life.