A restrictive ventilatory pattern is common in patients with univentricular heart after Fontan palliation and associated with a reduced exercise capacity and quality of life.

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The common restrictive ventilatory pattern in patients with a Fontan circulation has a substantial impact on exercise capacity and life’s quality. Associated risk factors are diaphragmatic paralysis, scoliosis, a high total number of interventions, and cachexia.

BACKGROUND

The Fontan circulation is highly dependent on ventilation, improving pulmonary blood flow and cardiac output. A reduced ventilatory function was reported in these patients, but its extent and consequences are unknown.

OBJECTIVES

This study investigates the extent and characteristics of the ventilatory impairment in Fontan palliated patients, its predictors and subsequent outcomes.

METHODS AND PATIENTS

This European multicenter retrospective cross-sectional study included 232 patients (140 females, 92 males, age 25.6±10.8). In mean 21.0±7.0 years after Fontan surgery. Resting spirometry, cardiopulmonary exercise tests, and health related self-perceived quality of life assessment (SF-36) were performed from 2003 to 2015.

RESULTS

VENTILATORY FUNCTION and SPIROMETRIC TESTS

Independent predictors of FEV₁

Results of a multiple regression analysis

DIAPHRAGMATIC PARALYSIS p=0.01
SCOLIOSIS p=0.001
TOTAL NUMBER OF INTERVENTIONS p=0.002
BMI < 18 p=0.01
PROTEIN LOOSING ENTEROPATHY p=0.02

VENTILATORY FUNCTION and EXERCISE CAPACITY

A normal FEV₁ was associated to better results in the CPET. The group of patients with a restrictive pattern had a peakVO₂%pred. of 61.98±15.71 versus the 73.50±17.60% of those with a preserved ventilation, setting a high significant difference (p<0.0001). This happened independently from sex, age, BMI, years from Fontan, total number of interventions, scoliosis, diaphragmatic paralysis, PLE, and NT-pro-BNP.

VENTILATORY FUNCTION and QUALITY OF LIFE

The mean scores of the SF-36 questionnaire were significantly reduced, compared to the expected values, in the categories of physical functioning (p=0.001), physical role functioning (p=0.005), general health (p=0.001), vitality (p=0.02), social role functioning (p=0.01), emotional role functioning (p=0.001), and health transition (p=0.02).

CONCLUSIONS

There is a dramatically high prevalence of restrictive ventilatory pattern in Fontan palliated patients. The associated predictors are:

- Total number of interventions
- Long-term complications
- Cachexia
- FEV₁ directly and independently correlates with:
  - Exercise capacity
  - Quality of life

1. The spirometric analysis should be integrated in their follow-up;
2. The associated risk factors should be prevented and aggressively treated;
3. Methods to improve ventilatory functions should be tested prospectively;

STUDY LIMITATIONS

- No body plethysmography was available;
- Only punctal data were considered;
- The early development factors were not analyzed;

No disclosure - comments to alessiacallegari@gmail.com