Cardiac function associated with home-ventilator care in Duchenne muscular dystrophy

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Background: Duchenne muscular dystrophy (DMD) is a prevalent neuromuscular disorder that affects one in 3600 live male births. Most leading cause of mortality in DMD patients is usually respiratory complications or cardiomyopathy. But nowadays, cardiomyopathy is becoming the leading cause of death in DMD patients because mechanically assisted lung ventilation and assisted coughing help resolving respiratory complication. We investigated to reveal the relation of cardio-pulmonary function in DMD patients.

Method: We retrospectively reviewed the DMD patients diagnosed from 2010 January to 2016 March in Gangnam Severance Hospital. Demographic characteristics, pulmonary function factor, Echocardiography data were assessed.

Results: We reviewed 54 case of DMD and divided into two groups (Home-ventilator assisted group vs Non-ventilator assisted group). In pulmonary function analysis, Home-ventilator group (1038±620.41ml) show lower sitting Functional vital capacity than Non-ventilator group (1455±603.12ml). In echocardiographic measurement, mean LV Ejection fraction and Fractional shortening were higher in Home-ventilator group. Home-ventilator assisted group’s E/A ratio (1.7±0.44) was more stable than Non-ventilator assisted group (2.02±0.62). With estimation by the multiple linear regression test, TDI S’ was higher in Home-ventilator group (Estimated β : 1.06, Standard error : 0.48).

Conclusion: DMD patients with ventilator care might have better systolic and diastolic cardiac function. Especially, patients without ventilator assistance may need more meticulous evaluation for cardio-pulmonary function, which may help to care the early cardiopulmonary dysfunction.