Is the 6 minute walk test or platelet count helpful in determining prognosis of patients with Eisenmenger Syndrome and Down syndrome?

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Introduction. 6MWD (6 minute walk test distance) is the standard test in the clinic for patients with Pulmonary hypertension. Doubt has been cast on its usefulness. We wished to try to determine whether this or blood tests were good measures of prognosis in our patients with Eisenmenger syndrome (ES) and Down syndrome (DS)

Methods. Patients with ES and DS had serial 6 MWD, oxygen saturations at rest and on exercise and full echocardiographic assessment from our pulmonary hypertension clinic in the last 3 years. RV function was indicated by TAPSE and lateral wall S'. Platelet count was corrected by haematocrit for the presence of erythrocytosis.

Results. Of 154 ES patients, 89 had DS (42 male) with mean resting saturations of 81%. WHO classification was a poor indicator of 6MWD (WHO I = 279m, WHO II = 279m, WHO III = 256m, WHO IV = 215m) and did not correlate well (coefficient 0.15). WHO class and 6MWD did not correlate overall with TAPSE or S' but within class III this became significant (correlation = 0.46, p=0.02). Saturation at start and end exercise did not correlate with 6MWD, survival or RV function. 79 DS patients were commenced on therapy (phosphodiesterase inhibitor or endothelin receptor antagonist) but the change in 6MWD was not significant. TAPSE and S' correlated well with each other, but only predicted 6MWD and survival in WHO class III. 55% of DS patients had a low platelet count <150^9/L (range 14-148) with a strong positive correlation between platelet count and rest oxygen saturations, even when corrected for Haematocrit. The lower counts were also strongly predictive of a poorer prognosis.

Conclusions. We did not find that the 6MWD or RV function was a good indicator of prognosis in DS unless within WHO group III. This suggests that until the condition is more severe, our measures of assessment are not sensitive enough to detect significant change. Platelet count is a predictor of poor prognosis, especially in DS patients, but only partly due to bone marrow hypoxia. Further measures are needed to allow better determination of prognosis in DS patients with Eisenmenger syndrome.