subgroups. Participants completed validated, age-appropriate questionnaires examining standard psychological parameters. Participants also underwent an evaluation of exercise, including formal exercise stress testing and measurement of free-living activity using an ActiGraph accelerometer. Results were analysed using parametric methods. 148 patients (mean age 15.6 years) consented to participate, 86 were male (60%) and 105 had major CHD (73%). Diagnostic subgroups included 59 acyanotic (27.3%), 61 acyanotic corrected (42.7%), 30 cyanotic corrected (21.0%) and 15 (9%) cyanotic palliated patients. Beck Youth Inventory demonstrated corrected (42.7%), 30 cyanotic corrected (21.0%) and 13 (9%) cyanotic subgroups included 39 acyanotic (27.3%), 61 acyanotic palliated patients, higher anxiety scores (p value 0.01 (~5.42, ~1.13)). There were no significant differences across study groups for self-esteem or other psychological parameters. 134 participants (95.7%) took part in regular exercise each week. There was no significant difference in activity score between study groups. On formal exercise testing, more complex patients performed worse at peak exercise. Exercise time for acyanotic group 11.73 mins (sd 5.74) compared to 8.26 mins (sd 4.08) in cyanotic palliated group, p value 0.002 (1.32, 5.61)). However, patients with major CHD had significantly higher activity counts. Correlation analysis showed that self-esteem and health locus of control were important predictor variables for activity. Self-esteem and mood seem well preserved in adolescents with CHD as a whole. The majority of young people with CHD, in this group, take part in regular exercise. Surprisingly, complex patients rate themselves to be as active as those with minor CHD. While accelerometer data indicate that the group may be more active day to day, they are limited in terms of peak exercise duration. The experience of growing up with a chronic condition may therefore have a positive effect on psychological health and interventions targeted around this area may influence activity.

**134 MUTATIONS IN THE SARCOMERE PROTEIN GENE MYH7 IN EBSTEIN’S ANOMALY**

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**Background** Ebstein’s anomaly is a rare congenital heart malformation characterised by adherence of the septal and posterior leaflets of the tricuspid valve to the underlying myocardium. As there have been reports of abnormal left ventricular morphology and function in patients with Ebstein’s anomaly we hypothesised that mutations in the β-myosin heavy chain (MYH7) may be associated with Ebstein’s anomaly.

**Methods** MYH7 mutation analysis was undertaken in 141 unrelated affected individuals with Ebstein’s anomaly using next-generation sequencing on the 454 platform. 64 probands had no associated cardiac anomalies. The most common associated cardiac malformations were atrial septal defect (48 probands) and left ventricular non-compaction (LVNC) (7 probands). Where mutations were discovered, family studies were undertaken and the segregation of the mutation with disease was investigated.

**Results** Heterozygous mutations were identified in eight of the probands including six of the seven with LVNC. Two patients had the same mutation; of the seven distinct mutations, five were novel (four missense changes and an in-frame deletion) and two have been previously reported in patients with hypertrophic cardiomyopathy. Family studies revealed additional members with LVNC for three of the probands, one of whom also had a relative with Ebstein’s anomaly. In these three pedigrees the mutation segregated with disease.

**Conclusions** Mutations in MYH7 occur relatively frequently in Ebstein’s anomaly accompanied by LVNC. This study is another example of mutations in a sarcomere protein causing congenital heart malformation.