Value of FOT in young children with SMA

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Body: Spinal muscular atrophy (SMA) causes respiratory compromise that is difficult to assess in young children. Forced Oscillation Technique (FOT) is commercially available for children as young as 2 years and doesn’t require forced expiration. Aim: To assess usefulness of FOT in young children with SMA. Methods: Children with SMA aged <10 years were recruited. FOT was performed monthly for 12 months (5 measurements). Spirometry, assisted and unassisted peak cough flow (PCF) were performed where possible. Height was predicted from ulna length. Predicted values and z-scores were calculated from height. The paired t test compared observed and predicted values. Regression analysis assessed relationships between FVC and PCF to FOT values. Clinical information included: SMA type, chest infection history, sleep study results, Cobb angle, Valproate use and mobility level. Relationships to FOT were sought using ANOVA. Results: 12 children (7 male) were recruited, mean age 6.26 (+2.59) years. $\text{Rrs}_8$ (mean z-score +2.04 ±2.22, p<0.001) and $\text{Xrs}_8$ (mean z-score -1.49 ±1.74, p=0.01) are significantly abnormal. 4 children performed spirometry. A linear relationship occurs between $\text{Xrs}_8$ and FVC ($R^2 0.54$), unassisted ($R^2 0.33$) and assisted ($R^2 0.43$) PCF. Over 12 months $\text{Xrs}_8$ z-score improved (rate of change of +0.44 z-score per annum p=0.001), $\text{Rrs}_8$ z-score worsened (rate of change +0.23 z-scores per annum p<0.02). No relationship (p>0.05) was found between clinical characteristics and FOT values. Conclusion: FOT testing is feasible in young children with SMA; with abnormal values of $\text{Xrs}_8$ and $\text{Rrs}_8$ on grouped data. $\text{Xrs}_8$ is related to respiratory tests used to monitor progress in SMA (FVC, PCF). Further research on the value of FOT in managing individuals is warranted.