

Increased Medical Interventions in Children with 22q11.2 Deletion Syndrome (Velocardiofacial Syndrome)

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2011

Abstract

Velocardiofacial syndrome (VCFS) or 22q11.2 deletion syndrome has a wide range of associated features including congenital heart defects, palate abnormalities, and learning disabilities. Medical interventions in infants and young children with VCFS have been described in the literature, but information diminishes as children reach school age. It was anticipated that children with VCFS would experience an increased number of surgical procedures and hospitalizations compared to children with isolated congenital heart defect (CHD), isolated cleft palate (CP), and children with no genetic syndromes or birth defects. To test this we conducted a retrospective medical record review of 77 children with VCFS, 35 children with CHD, 30 children with CP, and 32 from the local community. Numbers and types of surgical procedures and hospitalizations were compared between the groups using analysis of variance methods. Children with VCFS had a significantly greater number of surgical procedures (adjusted $p < 0.001$ each comparison) and hospitalizations (adjusted $p < 0.01$ each comparison) than children with CHD, CP, and the LC. Overall, 81% of the VCFS cohort had at least one surgical procedure and 53% had at least one hospitalization. Forty-five percent of children with VCFS and a palate defect had at least one surgical procedure on the palate between the ages of 5-10 compared to only 17% with CP. The results of this study indicate that children with VCFS had a significantly increased need for medical interventions between the ages of 5-10 compared to children with non-syndromic CP, CHD, and from the LC. Information from this study can be used to create management protocols for health care providers and to provide anticipatory guidance to caregivers of children with VCFS.