Closure of the Spheno-occipital Synchondrosis in Patients with Crouzon Syndrome: A Link to Midface Hypoplasia

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Purpose: Premature fusion of the spheno-occipital synchondrosis (SOS) has recently been described in the syndromic craniosynostosis subpopulations with Apert syndrome and Muenke syndrome. The current study aims to characterize SOS fusion in patients with Crouzon syndrome, a syndrome with high rates of midface hypoplasia.

Methods: A retrospective case-control study was performed in patients with Crouzon syndrome treated at a large craniofacial center between 1984 and 2012. Inclusion criteria were a diagnosis of Crouzon syndrome and at least one high-quality CT scan in which SOS patency could be assessed. Age/gender matched control CT scans were identified and assessed for status of SOS patency. Three independent reviewers with high inter-rater reliability (kappa=.88) graded SOS patency on axial images as open, partially fused, or completely fused SOS. Wilcoxon Rank-Sum test was used to compare the Pfeiffer group to controls.

Results: Over the study period, 30 patients were identified with Crouzon syndrome. A total of 24 patients with 112 head CT scans met inclusion criteria. All patients with Crouzon syndrome had some degree of midface hypoplasia. Accordingly, 112 age/gender matched control CT scans were assessed, and no patient in the control group had midface hypoplasia. Within the Crouzon group, the average age of complete closure (14.33 ± 3.43 years; n=31) evident on CT scan was significantly younger than the control group (16.55 ± 2.15 years; n=18) (p=.0155). The average age of partial closure evident on CT scan was significantly younger (5.57 ± 2.04 years; n=43) within the Crouzon group compared to the control group (10.65 ± 2.44 years; n=18) (p=.0001). The average age of scans showing complete patency of the SOS in the Crouzon group (1.32 ± 1.07 years; n=38) was significantly younger than the control group (3.22 ± 2.30; n=76) (p=.0001).

Conclusions: The SOS closes significantly earlier in patients with Crouzon syndrome compared to age/gender-matched controls. Although causality cannot be concluded, there exists a strong correlation between midface hypoplasia and premature SOS closure in Crouzon syndrome.