Long-term Follow-up of Pre-operative Infant Event-related Potential in Children with Craniosynostosis

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**Purpose:** Event-related potentials (ERPs) provide a real-time safe, noninvasive method to assess neural networks in infancy. Limited data exist, however, as to the predictive value of these results with eventual neurocognitive outcomes at school age. The purpose of this study was to investigate whether ERP recordings at infancy also translated into neurocognitive deficits at school age.

**Methods:** 16 infants who underwent evaluation of auditory processing in infancy were followed up for neurocognitive testing. ERPs were extracted from electroencephalographic data evoked in response to language stimuli, and were found to have a positive inflection peaking 150 to 300 msec (P150) after presentation of the stimuli, followed by a negative wave 300 to 700 msec (N450) after. The P150 and N450 components were examined over the right and left frontotemporal scalp areas. Patients underwent a battery of neurocognitive tests including assessments of IQ (Weschler Abbreviated Scale of Intelligence 2nd edition), academic achievement (Kaufman Test of Educational Achievement 3rd edition), and motor coordination (Beery-Buktenica Development Test of Visual-Motor Integration 6th edition). Absolute deviation of ERP results from age-matched controls were computed. Spearman’s correlations were used to assess the strength of correlation between neurocognitive scores and ERP values, as well as other sociodemographic variables.

**Results:** 11 patients with ERP in infancy were neurocognitively tested. Average time to follow-up neurocognitive testing was 10.1 ± 1.7 years. 7 (63.6%) were male, 9 (81.8%) were white. 7 (63.6%) underwent whole vault cranioplasty while 4 (36.4%) underwent endoscopic strip craniectomy. Mean age at surgery was 7.4±2.5 months. A greater absolute difference from age-matched controls in the left-sided P150 component was correlated with worse outcomes in the following domains: full-scale IQ (r=-0.709, p=0.015), performance IQ (r=-0.673, p=0.023) and fine motor skills (r=-0.654, p=0.029). While aberrancy in the P150 component was also correlated with math achievement (r=-0.065, p=0.028) and reading comprehension (r=-0.603, p=0.036), significant correlations were also found between math achievement and mother and father’s highest level of education, and between reading comprehension and mother’s highest level of education and household income. There was no correlation, however, between any demographic variables and full-scale IQ or performance IQ. Various socioeconomic factors were found to correlate with math and reading achievement, but not IQ or motor skills.

**Conclusion:** Brain activity measured at infancy has predictive value for eventual neurocognitive outcome, with greater deviation from normal being associated with greater IQ deficits. Early ERP assessment may help manage patient expectations about neurocognitive outcomes following intervention.

Prenatal Diagnostic of Pierre-Robin Sequence (PRS) Using Ultrasound

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**Purpose:** Pierre-Robin Sequence (PRS) is characterized by micrognathia and glossoptosis and can result in upper airway obstruction. Prenatal diagnosis of PRS facilitates delivery team preparation for an airway emergency. Routine 20-week ultrasounds screen facial features and can characterize mandibular morphology and the maxillomandibular relationship. This study aimed to evaluate 20-week fetal
ultrasounds to determine if specific facial measurements could predict PRS diagnosis and disease severity.

**Methods:** A retrospective case control study of 40 patients with PRS and 40 gender-matched controls (24 male, 16 female in each cohort) between January 2014 and May 2019 was performed. Respiratory and surgical interventions were recorded and scored for severity. Mid-sagittal profile images of the ultrasounds were reviewed and measured for 3 parameters to assess micrognathia: facial nasomental angle (FNMA), facial-maxillary angle (FMA), and alveolar overjet. FMA <66° and FNMA <136° signify micrognathia. A one-tailed t-test was calculated and the mean ± standard deviation was reported.

**Results:** Patients with PRS demonstrated significantly smaller FNMA compared to the control group (129.6 ± 9 vs.137.9 ± 2.8, respectively; *p*<.001). FMA was significantly smaller in the PRS group compared to the control group (64.1 ± 9.3 vs. 75.3 ± 6.5; *p*<.001). The PRS group also demonstrated significantly larger overjet compared to the control group (3.7 ± 1.3 vs. 2.3 ± 0.8, respectively, *p*<.001). In the PRS cases, 38% did not require external respiratory support (n=15), 43% needed supplemental oxygen by nasal cannula or CPAP (n=17), and 20% were intubated (n=8). As respiratory support needs increased, median FMA decreased and alveolar overjet increased. In the control group, 93% required no external respiratory support (n=37) and 7% needed supplemental oxygen by CPAP (n=3). In the PRS cases, 45% received mandibular distraction or tongue-lip adhesion (n=18), 15% underwent supraglottoplasty only (n=6), 8% required tracheostomy (n=3) and 32% did not require surgical intervention (n=13). Surgical patients tended to have smaller FNMAS and greater overjet compared to nonsurgical patients; median FNMA was 127° versus 132°, and median overjet was 2.8 versus 4.15 mm, respectively, however this was not significant. None of the control patients underwent surgical intervention.

**Conclusions:** Mandibular features on 20-week anatomy ultrasound can be measured to predict PRS prenatally and prepare for respiratory intervention at delivery to minimize hypoxia at birth. Alveolar overjet, previously not described in prenatal ultrasound literature but routinely assessed on neonatal clinical evaluation, is measurable and has utility in prenatal diagnosis, as do FMA and FNMA.

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**QS1**

**Nasopharyngeal Airway and Subcranial Space Analysis in Pfeiffer Syndrome**

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**Background:** Tracheotomy in infancy helps patients with Pfeiffer syndrome survive by preventing respiratory crisis, however, difficulty in decannulation consequently may be a challenge. This study attempts to investigate the regional abnormalities of the nasopharyngeal airway in Pfeiffer syndrome, to provide an anatomic basis for the upper airway surgical treatment and decannulation.

**Method:** Seventy-two preoperative CT scans (Pfeiffer syndrome, n=30; control, n=42) were included in this study. The airway volume, cross-sectional area, and cephalometrics, were measured using Materialise software.

**Results:** Patients with Pfeiffer syndrome developed a 50% (p<0.001) reduction of nasal airway volume, and a 44% (p=0.003) restriction in pharyngeal airway volume. The cross-sectional area of Pfeiffer patients at the choana, was only half that of normals’ (p<0.001). The posterior width of nasal airway in Pfeiffer syndrome was shortened by 13% (p=0.003), and the height was reduced by 21% (p<0.001). The cross-sectional areas at condylion and gonion levels, indicating the caliber of pharyngeal airway at entrance and midsection, were reduced by 67% (p<0.001) and 47% (p<0.001), respectively, when compared with normals.

**Conclusion:** Nasal airway volume in Pfeiffer syndrome is significantly restricted, in length, height and width, along with the choanal stenosis in all cases in this cohort. The reduced anteroposterior length of nasal airway contributes to the shortened maxilla, more than the anteroposterior...