

6-2014

# Neuropsychological Outcomes of Children with Craniosynostosis

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Running head: Neuropsychological testing and craniosynostosis

Neuropsychological Outcomes of Children with Craniosynostosis

By

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Senior Thesis

A thesis presented in partial fulfillment

of the requirements for the degree of

Bachelor of Science

Department of Psychology

UNION COLLEGE

Schenectady, New York

June 2014

ABSTRACT

STRAHLE, MARYKATHRYN Neuropsychological Testing on Pediatric Patients with Craniosynostosis.

Department of Psychology, June 2014.

ADVISOR: Cay Anderson-Hanley.

Craniosynostosis is the premature fusion of one or more cranial sutures. Previous research suggests that craniosynostosis is related to cognitive deficits, impairments, and delay, and that these impairments might be related to the age of surgical intervention or the location and severity of suture fusion (Kapp-Simon, 1998). The current research sought to further investigate such relationships using a sample of 177 pediatric patients from the University of Michigan Neuropsychology Clinic. The data included 56 intelligence quotient (IQ) scores from various neuropsychological tests [Bayley Scales of Infant Development (BSID), Wechsler Preschool and Primary Scale of Intelligence (WPPSI), and Wechsler Intelligence Scale for Children (WISC)]. Results indicate that when surgery is conducted before age two ( $n = 46$ ), post-surgical IQ is not related to age ( $r = ; p =$ ); however, patients who receive corrective surgery for craniosynostosis after two years of age appear to be at a higher risk for cognitive impairment.

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### Neuropsychological Testing on Pediatric Patients with Craniosynostosis

Infancy is one of the most important and critical stages of life, and what happens to the human body during infancy can affect an individual's entire trajectory of development. Infants grow a great deal during their first years of life, but both the brain and skull specifically undergo significant and intense growth. The proper growth and development of the brain and skull is imperative for the subsequent development of the rest of the body. Any malformations or defects of the brain and skull that present during infancy can cause serious damage or delay (Speltz, Kapp-Simon, Cunningham, Marsh, & Dawson, 2004).

Craniosynostosis is one such malformation that can present during infancy, and is described as the premature fusion or ossification of one or more cranial sutures. The fusion of at least a single suture occurs in one of 2,000 live births and affects males three to four times more than females (Speltz et al., 2004; Boltshauser, Ludwig, Dietrich, & Landolt, 2003). The sagittal, metopic, coronal, and lambdoid are the four primary sutures investigated in the cases of craniosynostosis, with sagittal and metopic being the two most common. The condition can manifest as either non-syndromic or syndromic (e.g., Apert, Crouzon, Pfeiffer, or Saethre-Chotzen).

The current literature investigates the patterns, implications, and treatment of craniosynostosis. Researchers also study the effect craniosynostosis has on cognitive development and executive function; especially the role specific suture fusions might have on certain cognitive functions. Individuals in this field have suggested that future studies examine both the role of surgical intervention as well as the timing of surgical treatment. Overall, scholars

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who research craniosynostosis seek solutions as to how to best treat individuals with these skull malformations.

### *Potential Effects of Craniosynostosis on Cognitive Function*

For about three decades, studies have investigated cognitive impairments in individuals with craniosynostosis. However, results have differed in their interpretations and implications, and there are different speculations as to whether craniosynostosis truly affects cognition in a negative way. Kapp-Simon (1998) researched the potential consequences of craniosynostosis regarding surgical correction and the presence of learning disorders. She predicted that surgical correction before one year of age would result in improved mental development scores, and that the commonness of learning disorders would be higher in untreated craniosynostosis patients. After a longitudinal study assessing patients at eight months, 21 months, and 50 months ( $n = 84$ ), the results demonstrated that surgical correction had no bearing on mental developmental scores in infancy. However, results from Renier, Brunet, and Marchac (1987) showed that 88 percent of children ( $n = ?$ ) who received surgery before one year of age scored a developmental quotient (DQ) of 90, while only 76 percent of children receiving surgery after one year of age had a similar level of function?. Such speculation of mental development scores in craniosynostosis patients has been ongoing and debated through dozens of studies. Therefore, different implications from different studies are not uncommon.

In addition to investigation of surgical intervention, researchers have begun to analyze the presence and location of suture fusion. Specifically, they have examined suture fusion in relationship to diagnoses of cognitive abnormalities like behavior, motor skills, speech and language. Becker et al. (2005) examined the presence of such abnormalities in a total of 214 patients with various types of craniosynostosis. Cognitively, the results showed 45 percent of

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patients exhibited psychological testing abnormalities. Patients were considered abnormal if they scored at least one standard deviation below the mean on both behavioral and cognitive psychological tests. Additionally, Becker et al. found that speech abnormalities were present in 23 percent of patients. These results suggest that there is reason to investigate cognitive impairments in craniosynostosis patients, but the findings do not offer any convincing conclusions.

Toth et al. (2008) also investigated the relationship between suture fusion and cognitive abnormalities and impairment with cognitive tests. Through a developmental lens, the study examined whether “the global delays found among infants and toddlers with single-suture craniosynostosis (SSC) may develop into more specific forms of impairments in later years” (2008). Comparing 200 toddlers with surgically corrected SSC to 149 unaffected toddlers over time, the researchers found that the two groups performed almost equally on the AB and ABID tasks.

While many researchers have examined the potential effects of craniosynostosis with the use of professionally administered standardized tests, Knudsen and colleagues (2012) used parental questionnaires. The purpose was to determine whether this method could uncover any psychological malformations via parental knowledge and perception of children. Knudsen et al. (2012) yielded results consistent with previous literature in that patients who received surgical intervention did not demonstrate psychological impairment pre-surgery or post-surgery, but arrived as such results through the estimation of patients’ parents. This study is imperative for the future of craniosynostosis treatment because any exiting data available for craniosynostosis patients can help better the treatment and prognosis for craniosynostosis patients.

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### *Sagittal Synostosis*

Sagittal synostosis occurs when the sagittal cranial suture ossifies prematurely. This is the most common form of synostosis, occurring in one out of 5,000 live births (Kapp-Simon, Speltz, Cunningham, Patel, Tomita, 2007). The sagittal suture runs down the middle of the skull from the anterior to the posterior [Image 1.1]. When it fuses, the skull appears to narrow in width and extend in length, resulting in an appearance clinically referred to as scapholcephaly (Kapp-Simon et al., 2007).

Families of children with craniosynostosis often consider surgical reconstruction of the skull, and the effects and consequences of surgical correction have been investigated. Some implications of surgical intervention have been believed to be beneficial, and thus the questions remains as to whether a lack of surgical intervention will be harmful. Because of this, researchers have conducted studies investigating the lack of surgery on craniosynostosis patients.

Boltshauser et al. (2003) evaluated 30 patents with isolated sagittal craniosynostosis (ISC) an average of 9.25 years after they had received an evaluation during infancy. They examined head shape, behavior, school performance, quality of life, and administered neuropsychological tests. Out of the 30 patients, 17 had siblings used as controls. The assessments yielded lower scores of positive emotions, but still psychological adjustment within a normal range. Additionally, intellectual performances of these patients were also within an average range.

These findings suggest that patients who do not undergo surgical correction of their craniosynostosis, at least sagittal, are not at an increased risk for intellectual impairment. The research of Shipster et al. (2003) supplements the findings of Boltshauser et al. (2003), by investigating language, speech, and cognitive impairment in 76 patients with isolated sagittal synostosis (ISS). The patients, aged 9 months to 15 years 7 months, showed no increased



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cognitive impairment. However, amongst 37 percent of the sample there was a high prevalence rate of speech and/or language impairment. This suggests that sagittal synostosis might not affect cognitive functioning, but perhaps speech and language functioning specifically.

Therefore, research regarding deficits unique to specific locations of suture fusions is imperative to the craniosynostosis field. The results of Shipster et al. (2003) suggest that perhaps language and speech deficits are prevalent in sagittal synostosis patients. However, it is unclear as to whether these deficits are unique to sagittal synostosis, or are results of craniosynostosis at all. Ruiz-Correa et al. (2007) examined another potential effect of sagittal suture synostosis, investigating whether the severity of the scaphocephalic malformation affects neurodevelopment. The researchers evaluated the neurocognitive development of 75 infants, with a median age of 4.5 months. Based on evaluations using the mental (MDI) and motor (PDI) scales of the Bayley Scales of Infant Development and the Preschool Language Scale, the results showed no relationship between neurodevelopmental status and the sagittal shape of the skull, no matter how severe.

The research on sagittal synostosis, the most common form of craniosynostosis, presents questions that have yet to be answered. It is unclear as to whether the uniqueness of the sagittal suture causes specific, if any, cognitive impairment. Prior literature suggests that internalizing symptoms may be present in sagittal synostosis patients (Boltshauser et al., 2003), in addition to speech and language deficits (Shipster et al., 2003). However, these studies further suggest that intellectual functioning is not affected significantly. While sagittal synostosis is the most common, the second most common synostosis is fusion of the metopic suture. Metopic synostosis also has been investigated in terms of its location and potential deficits unique to this synostosis. By comparing the findings of metopic synostosis research to those of sagittal

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synostosis, we may further be able to determine the characteristics and implications of craniosynostosis in more meaningful detail.

### *Metopic Synostosis*

Metopic craniosynostosis occurs in approximately 1 in 15,000 live births (Starr et al., 2010). The metopic suture is located at the anterior of the skull, and crosses the skull laterally [Image 1.1]. The presenting symptom of metopic synostosis is trigonocephaly, resulting in a triangular and pointed forehead. Researchers have investigated cognitive impairment, behavioral, developmental, educational, and motor problems, as well as speech and language. Furthermore, some studies have examined the relationship between the severity of trigonocephaly on cognitive development, as the study of Ruiz-Correra et al. (2007) did with scaphocephaly.

Researchers Sidoti, Marsh, Marty-Grames, and Noetzel (1996) investigated the relationship of metopic synostosis and cognitive impairment as well as behavioral disturbances. By examining 32 patients with both prospective data (questionnaires) and retrospective data (craniofacial evaluations), Sidoti et al. discovered that more than one third of patients exhibited behavioral or cognitive abnormalities. However, not all patients had data for each type of data collection. For example, not all patients had retrospective evaluations, and others were unable to be contacted for follow-up questionnaires. It is often difficult to garner a sample complete with data for all analytic measures pertinent to the study.

While Sidoti et al. (1996) examined behavioral disturbances and cognitive impairment in one sample, Kelleher et al. (2006) investigated behavioral disturbances alongside developmental and educational problems in children with metopic synostosis. They observed a sample of 63 children, 30 percent of patients presented with mild trigonocephaly and were treated without

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surgical correction. The remaining percentage of patients underwent surgical correction at an average age of one year.

Parents of children with metopic synostosis completed a questionnaire that sought to determine whether a child experienced behavioral or developmental difficulties, as well as difficulties in school. Parental reports indicated that 33 percent of children showed developmental delay, and 37 percent of parents were concerned with their child's behavior. Last, out of the 42 children who attended school, 10 percent in a mainstream school setting repeated one year. These statistics suggest that deficits and abnormalities are not inescapable, but that they are definitely not guaranteed. Importantly and most conclusively, Kelleher et al. determined that there was no statistically significant difference between patients who received surgery and those who did not in the developmental, educational, or behavioral domains (2006).

There was a number of patients in the study of Kelleher et al. (2006) who were evaluated for developmental problems and who did not receive surgical treatment. Warschausky et al. (2004) investigated development in a similar sample of patients. The researchers conducted a retrospective cross-sectional study, with a sample of 22 infants with metopic synostosis before they underwent surgery. The patients were evaluated for developmental delays, specifically in cognitive, motor, and language domains using the Bayley Scales of Infant Development (BSID). The study could not conclude with an association between metopic craniosynostosis and lower motor and cognitive development. However, results did suggest that children with metopic synostosis might demonstrate slower development of language acquisition.

Speech and language development are important to investigate in patients with craniosynostosis, because the development of these skills occurs during a crucial imprinting period during infancy and toddlerhood. Therefore, the imperative question is whether metopic

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synostosis will compromise speech and language, but a second question asks whether the severity of presenting trigonocephaly affects speech and language development. Mendonca et al. (2009) evaluated twenty patients with metopic synostosis for language and speech delays. By use of five different measurements of the skull pre-operatively, a craniofacial team determined the severity of the metopic synostosis through computed tomography scans. All of the patients received corrective surgery at an average age of 1.25 years, and subsequently received a speech and language assessment at ages 3 and 5. Results demonstrated that six of the 20 patients showed developmental delay in speech and language, however the researchers discuss that these impairments cannot be attributed solely to the physical metopic severity.

Examining more general cognitive development, Starr et al. (2010) assessed the relationship between metopic severity and neurodevelopmental test scores. The metopic severity of 65 infants was determined before they underwent corrective surgery. Each patient was administered the Bayley Scales of Infant Development (BSID) as well as the Preschool Language Scale (PLS). Finding little evidence of an association between metopic severity and neurodevelopment, Starr et al. suggest that perhaps such findings would require a larger sample or different measurements of severity.

Craniosynostosis is a relatively new and under-researched field of study, and just as Starr and colleagues discuss, no data set is perfect and replete with all necessary data. Furthermore, craniosynostosis is a complex condition in which each case is unique. Generalizations are seldom made for cases of craniosynostosis, for each patient has her own trajectory in terms of confronting the condition. There are two complexities of craniosynostosis that have the ability to make craniosynostosis more severe and can greatly affect the prognosis. Syndromic craniosynostosis presents alongside other congenital malformations and deficits. Multi-suture

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craniosynostosis presents as a combination of two or more fused sutures, or in the case of pansynostosis for which every suture is fused.

### *Syndromic Craniosynostosis and Multi-Suture Craniosynostosis*

Syndromic craniosynostosis occurs “in conjunction with other anomalies that make up clinically recognized syndromes” (Derderian & Seaward, 2012). Apert, Crouzon, Saethre-Chotzen, and Pfeiffer are four of the most common syndromic craniosynostoses. Other syndromic craniosynostoses, like Robert’s, luckenschadel, frontonasal dysplasia, and kleeblattschadel are less common but still require complex treatment and medical attention. Individuals with syndromic craniosynostosis live with compromised skull development, increased risk for intracranial pressure (pressure buildup within the skull), exophthalmos (protrusion of the eyes), mid face hypoplasia (underdevelopment) and limb abnormalities (Derderian & Seward, 2012). Additionally, Chiari malformations can accompany syndromic craniosynostosis, which can eventually lead to hydrocephalus. These sequelae of syndromic craniosynostosis strongly influence the course of treatment and are more complicated than non-syndromic craniosynostosis. Since the brain reaches two-thirds its adult size within the first year of life, syndromic craniosynostosis severely inhibits proper skull growth and development more so than non-syndromic craniosynostosis.

Multi-suture craniosynostosis, is the fusion of two or more cranial sutures without any syndromic characteristics. However, multi-suture craniosynostosis is still more severe than single-suture craniosynostosis, and intracranial pressure is likely to be higher in multi-suture patients. Treatment, though, is extremely critical for all types of craniosynostosis. Some types of treatment include surgical intervention, while other methods are less invasive. Currently,

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researchers and craniofacial teams are striving to construct diagnostic and prognostic parameters for craniosynostosis treatment.

### *Current Treatment of Craniosynostosis*

The current treatments for patients with all types of craniosynostosis are varied and inconsistent. There are many factors that must be considered and addressed for craniosynostosis patients. These include obvious medical concerns, such as intracranial pressure and hypoplasia. Then, doctors must determine whether surgical correction is necessary, either for aesthetic reasons or to alleviate pressure caused by the malformed skull. Finally, depending on social factors such as age and socioeconomic status, health care providers must address any compromise in psychological well-being.

Addressing the lack of parameters in craniosynostosis evaluation and treatment, Ursitti et al. (2011) aim to present a holistic diagnostic strategy when treating patients with craniosynostosis. In patients with non-syndromic craniosynostosis, they state that a “careful physical, ophthalmological, and neurological examination is fundamental.” They suggest that perhaps current treatment of craniosynostosis is too focused on computed tomography (CT) scans and magnetic resonance imaging (MRI), and that these measures should be primary for syndromic craniosynostosis or cases in which the diagnosis is unclear (2011). Ursitti et al. (2011) recommend that pediatricians first determine the type of craniosynostosis, and secondly distinguish between the types that do and do not warrant surgical correction, lastly following with physical, ophthalmological, and neurological examinations.

However, Ursitti et al. (2011) overlook a facet of evaluation that provides invaluable information necessary for the most optimal treatment. Patient-reported outcomes, as suggested

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by Szpalski, Weichmann, Sagebin, and Warren (2011) enhance the quality and type of treatment. By understanding the patient's feelings and perspectives, craniofacial teams can provide more appropriate care. Including patient-report measures, Szpalski et al. (2011) generated a list of parameters for evaluation, diagnosis, and treatment for patients with craniosynostosis. Measures also include radiographic evaluations, neuropsychological assessments, and quality of life assessments.

Unfortunately, many craniosynostosis patients receive initial evaluations during infancy and therefore cannot provide (verbal) feedback. Additionally, since there are no set parameters for craniosynostosis treatment, options are slim and might default to surgical intervention. Unclear MRI or CT scans, ambiguous presenting symptoms, or even anxious parents could all be reasons surgery is performed in cases where it is not necessary. When surgery is warranted, however, correction and remodeling afford craniosynostosis patients an opportunity to live a life without the condition.

### *Surgical Intervention*

According to Lekovic, Bristol, and ReKate (2004), "Craniosynostosis remains primarily a surgical disease, the treatment of which is surgical release and cranial remodeling." They discuss that the implications of surgery include restructuring the skull as well as attempting to minimize cognitive deficits that craniosynostosis may potentially cause. However, the effects of surgical intervention as well as its timing remains disputed. Kelleher et al. (2006) note that, "it has been suggested that early surgical release of craniosynostosis might increase mental abilities." It is a widespread theory within the craniosynostosis field that early surgical intervention leaves the patient devoid of cognitive impairments that could have presented had surgery not been

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performed. While surgery does have evident aesthetic benefits, the claim for cognitive benefit does not have evidentiary support.

To determine whether or not untreated single-suture craniosynostosis (SSC) is associated with increased developmental delay, Da Costa et al. (2012) assessed 56 patients using the Bayley Scales of Infant Development (BSID). The patients ranged in age from 4 to 16 months and all were awaiting surgery. The results suggested that untreated SSC is in fact associated with developmental delay, specifically for patients in early infancy before receiving surgery. However, this study lacked post-operative data and thus an assessment of post-surgical development.

Analyzing a sample of 220 patients who had received both preoperative and postoperative cognitive assessment, Mathijssen, Arnause, Lajeunie, Marcha, and Renier (2006) investigated the effects of surgical timing. Patients had received either early surgical correction (less than one year of age) or late surgical correction (one year of age or older). The intelligence quotient (IQ) was acquired for each patient by the same team of psychologists. Comparisons were drawn between postoperative cognitive statuses of patients who had received surgery before one year of age and those who received it at or after one year of age. Mathijssen et al. (2006) found that there was no statistically significant difference between pre- and post-operative IQ scores.

### *Summary*

Therefore, the current literature investigating the role and effect of surgical intervention is inconsistent. Additionally, much emphasis is placed on the timing of surgical intervention, but



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it is unclear as to whether or not age at surgery affects cognitive abilities. Last, it is unknown as to whether specific types of craniosynostosis cause certain cognitive impairments.

### *Hypotheses*

Based on previous craniosynostosis literature and findings, it is hypothesized that:

1. Patients who receive surgery under the age of two years will have higher IQ scores than who received surgery older the age of two years (based on the results of Kapp-Simon, 1998, who investigated surgical patients under and over the age of one year).
2. Those with sagittal synostosis, the most common form of craniosynostosis, experience an increase of internalizing symptoms as well as language deficits (Boltshauser et al., 2003; Shipster et al., 2003).
3. Those with metopic synostosis, the second most common form of craniosynostosis, experience delayed motor skills, language deficits, and a higher incidence of behavioral problems (Sidoti et al., 1996; Warschausky et al., 2004).

## METHODS

### *Participants*

The sample (n=177) consisted of patients who were seen for assessment at the University of Michigan neuropsychology clinic, and contained 114 males and 63 females. Eighty-seven percent of the patients were Caucasian. All patients had received surgical intervention for their craniosynostosis between the ages 2.04 months to 9 years 2.52 months ( $M = 1.34$  years;  $SD = 1.46$ ; see Figure 2). Out of 177 patients, 153 had single suture craniosynostosis, and 24 had multi-suture craniosynostosis. Sixty-three single suture craniosynostosis patients presented with sagittal synostosis, 52 with metopic, 16 with bicoronal, 13 with left coronal, 9 with right coronal, and one with lambdoid (see Figure 3.1 and Figure 3.2). Twenty-nine patients presented with syndromic craniosynostosis (see Figure 4.1 and Figure 4.2).

### *Procedures*

The University of Michigan Neuropsychology Department ordered copies of neuropsychological test results available for all craniosynostosis patients who visited the clinic. The data of nine neuropsychological tests were catalogued in Microsoft Excel where it was available, but no data from three of the tests were yet analyzed in this report: Delis-Kaplan Executive Functioning System (D-KEFS), California Verbal Learning Test (CVLT), and the Trail Making Test (TMT).

Due to the retrospective nature of the study, no patient had data for each of the nine tests. Some patients had been administered the same test over more than one period of time, and all data were accounted for in the database. During data analysis, IQ scores from the Bayley Scales of Infant

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Development (BSID), Wechsler Preschool and Primary Scale of Intelligence (WPPSI), and Wechsler Intelligence Scale for Children (WISC) were used for pre-surgical and postsurgical test scores. If a patient had more than one IQ score at pre-surgery or post-surgery available, the score obtained at the youngest age was catalogued.

### *Measures*

Six neuropsychological tests were used in the present craniosynostosis data analysis to assess specific cognitive functions (see Table 1).

*Bayley Scales of Infant Development* (BSID; Bayley, 1993). The BSID is used to determine whether a child experiences delay in motor skills or cognition. It is comprised of the Mental Scale, Motor Scale, and Behavior Rating Scale.

*Mullen Scales of Early Learning* (MSEL; Mullen, 1995). MSEL evaluates cognitive ability and motor development.

*Wechsler Preschool and Primary Scale of Intelligence* (WPPSI; Wechsler, 1967). The WPPSI measures intelligence quotients in younger children.

*Wechsler Intelligence Scale for Children* (WISC; Wechsler, 1949). The WISC measures intelligence quotients in older children.

*Behavior Assessment System for Children* (BASC; Reynolds & Kamphaus, 2004). Parents, teachers, and patients themselves assess behavior by reading statements and indicating on a scale how relevant the statement is to the child on the BASC.

*A Developmental NEuroPSYchological Assessment* (NEPSY; Korkman, Kirk & Kemp, 1998). The NEPSY aims to assess a number of cognitive domains: attention and executive

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functioning, language, memory and learning, sensorimotor, social perception, and visuospatial processing.

### *Statistical Analysis*

The data collected was analyzed using Microsoft Excel and the Statistical Package for the Social Sciences (SPSS). Correlations and partial correlations were conducted in order to assess and associations between age and cognitive functions as well as locations and cognitive functions specified in the hypotheses above. T-tests and ANOVAs tests analyzed between-groups data.

## RESULTS

We first assessed the extent to which age at surgery predicted post-surgical IQ score. Out of the total 177 craniosynostosis patients in the database, 56 had available post-surgical IQ scores. These scores were obtained from Wechsler Preschool and Primary Scale of Intelligence, Wechsler Scale of Intelligence for Children or Bayley Scales of Infant Development depending upon the available data for each patient. In the instance that a patient had more than one IQ score, we chose to include the score obtained closest to the surgery in the analysis. The analyses were controlled for socioeconomic status (SES), but SES was available for certain patients ( $n = 91$ ) so the tradeoff with statistical power was balanced by running tests both with and without.

There was a negative association between age at surgery and post-surgical IQ scores ( $n = 56$ ;  $r = -0.48$ ,  $p = .002$ ), such that the younger a patient was when she received surgery, the higher she scored on a post-surgical IQ test (see Graph 1). However, when we assessed the same relationship only with patients who had received surgery under the age of 2 years, there was no association ( $n = 46$ ;  $r = -0.13$ ,  $p = .48$ ), such that there was no apparent effect on the post-

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surgical IQ scores for individuals who received surgery between 0 and 24 months of age (see Graph 2).

Then, we attempted to evaluate whether the type of craniosynostosis had an association with specific cognitive functions. The size of the sub-samples were not great enough to generate results for ANOVA tests, such that not enough sagittal synostosis patients had post-surgical data for internalizing symptoms and language measures (see Table 2), and not enough metopic synostosis patients had post-surgical data for behavior, motor, and language measures. (see Table 3).

## DISCUSSION

It has been speculated that craniosynostosis might contribute to cognitive impairment, deficit, and delay. Craniofacial teams of doctors and psychologists have investigated as to whether or not surgical intervention will prevent children from developing such cognitive impairments. However, research has demonstrated that surgical correction has no negative bearing on mental development scores in infants (Kapp-Simon, 1998) and toddlers (Toth et al., 2008). Furthermore, studies have researched whether or not the location of suture fusion affects specific cognitive domains. Potential deficits in language and increased internalizing symptoms have been investigated in patients with sagittal synostosis (Boltshauser et al., 2003; Shipster et al, 2003). Additionally, metopic synostosis and its relationship to behavior (Sidoti et al., 1996), and motor and language deficits (Warschausky et al., 2005) have been researched. The previous literature remains unclear and findings demonstrating significant relationship have not been replicated.

The current research sought to further investigate the questions posed by the previous craniosynostosis literature. This was achieved by obtaining and collating data from the

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neuropsychological tests administered to craniosynostosis patients at the University of Michigan Neuropsychology Clinic. We hypothesized that individuals post surgical IQ scores would be related to the age at which a patient received corrective surgery. Results indicated that there was a relationship when all patients with post-surgical IQ scores were included in the correlational analysis. However, the relationship became insignificant and nonexistent when individuals older than the age of two were removed from analysis. Secondly, we hypothesized that based on the previous literature; sagittal synostosis would have negative effects on internalizing symptoms and language. Also, we hypothesized that metopic synostosis would lead to behavioral delays, and motor and language deficits. These hypotheses were neither supported nor disproved: the sub-samples were not greater than ten patients (n=10).

### *Strengths*

Craniosynostosis research is imperative for the field of craniosynostosis treatment and prognosis. Since the condition is rare and under-researched, each patient and his or her health information are invaluable for research. This data set was unique in that it is one of the largest collections of pediatric patients with craniosynostosis. Furthermore, the information provided by neuropsychological tests is extensive and meaningful for psychological research.

### *Limitations*

As with any retrospective study, it is impossible to control the conditions each patient experiences. As was the limitation in gathering analyses for the effects of sagittal and metopic synostosis, and even post-surgical IQ scores, not every patient had test data for the same neuropsychological tests. Therefore, the sample size and statistical power was inevitably

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decreased. Additionally, not every patient had the same background information, such as socioeconomic status or genetic history. If the patient did have such data available, the data were difficult to find within patient records and at times were unclear.

### *Future Research*

Future research might seek to administer a uniform set of neuropsychological tests to each patient presenting to a clinic and agreeing to participate. A questionnaire should also be administered to each patient/patient family, so that demographics and genetics can be accounted for legitimately. This information can provide additional insight into any patterns present in craniosynostosis patients, and uniform assessment will make identifying such patterns easier.

, contributors to the field of craniosynostosis should strive to continue researching to determine appropriate treatments for craniosynostosis patients. The questions still remain as to whether or not surgery is purely aesthetic, or whether it will improve cognitive functioning potentially compromised by this condition. Furthermore, researchers should focus on patients with craniosynostosis who have not received surgery until after toddlerhood, and see if there is a common functionality of such patients. As in the current study, there was a group of patients who had received surgery late, much older than the age of two. These outliers presented a strong negative correlation between age at surgery and postsurgical IQ scores. However, when they were removed, the correlation was weak and not significant. This suggests that perhaps individuals have comorbidities that are severe, and thus must be addressed and treated before craniosynostosis.

As with any health condition, early intervention can have many benefits. Patients with craniosynostosis and their families should be vigilant and attentive throughout diagnosis and

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potential treatment. Researchers should continue assessing, evaluating, and investigating patients with craniosynostosis, in hopes of discovering patterns or links that can make the condition less severe.



ACKNOWLEDGEMENTS

1. Many thanks to my advisor, Professor Cay Anderson-Hanley, PhD., for all of her help, assistance and guidance.
2. Thank you to the University of Michigan Neuropsychology Department for permission to obtain neuropsychological test results of craniosynostosis patients.
3. Thank you to Dr. Jennifer Strahle, M.D., and Dr. Karin Muraszko, M.D., neurosurgeons at the University of Michigan, for permission to use this data set.
4. Many thanks to Dr. Mary Morris, PhD for her time and expertise in pediatric neuropsychology.
5. Thank you to the Student Research Grant (SRG) funding of Union College for making travel to University of Michigan possible.

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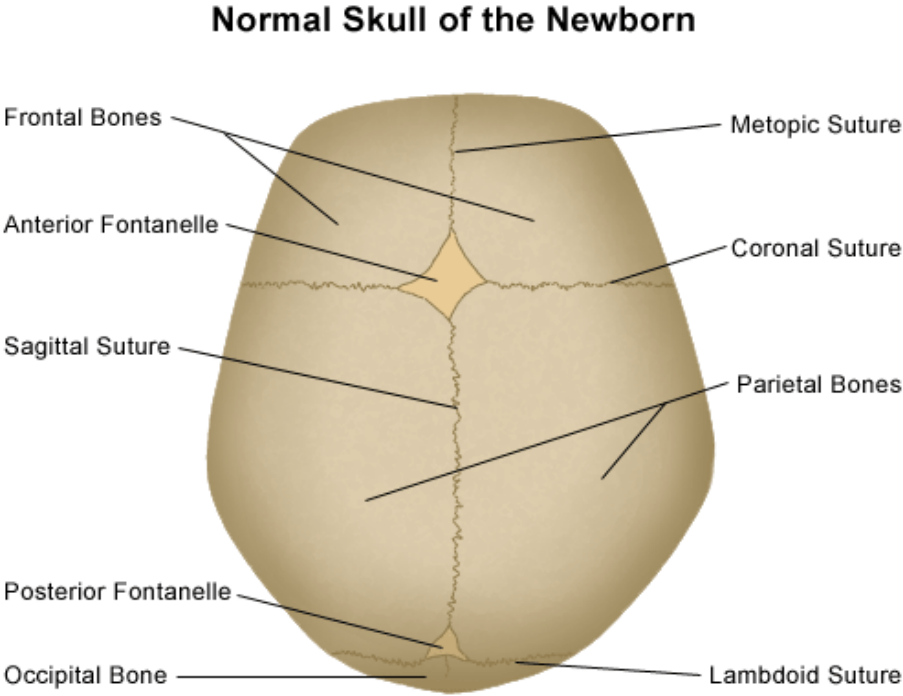


Figure 1. *The Normal Skull of the Newborn* (Stanford Children’s Health, 2014).

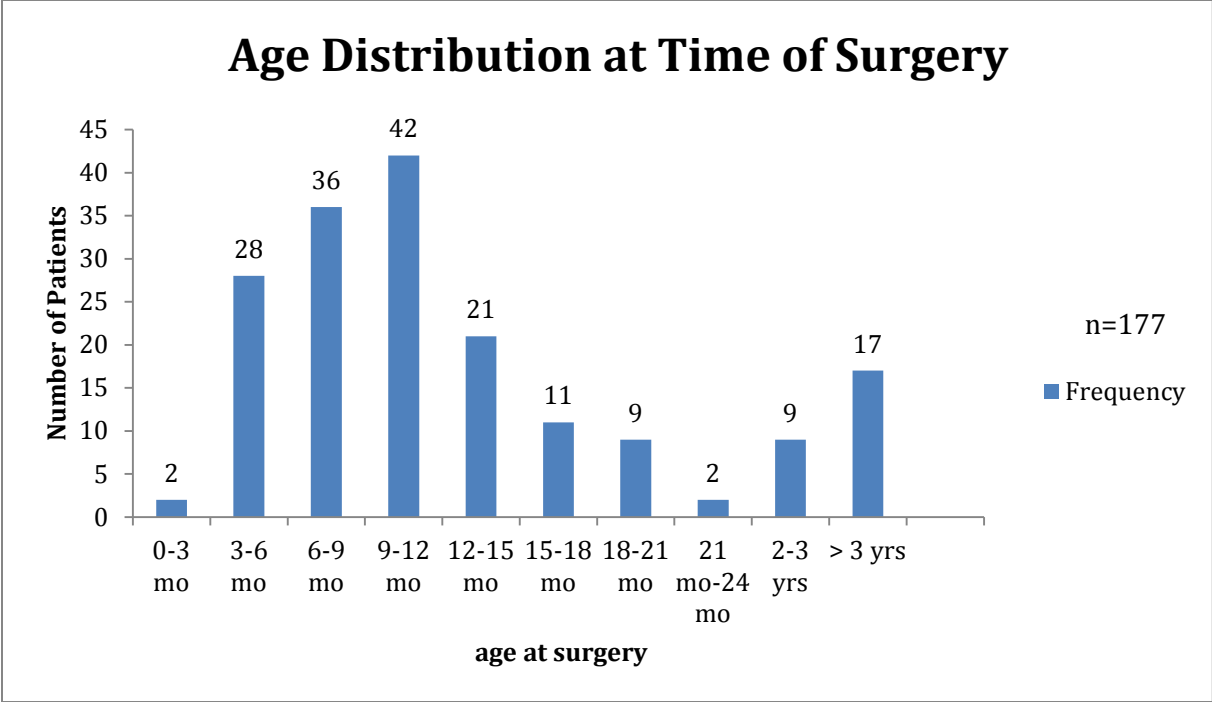


Figure 2. The age distribution of patients at the time they received corrective surgery.

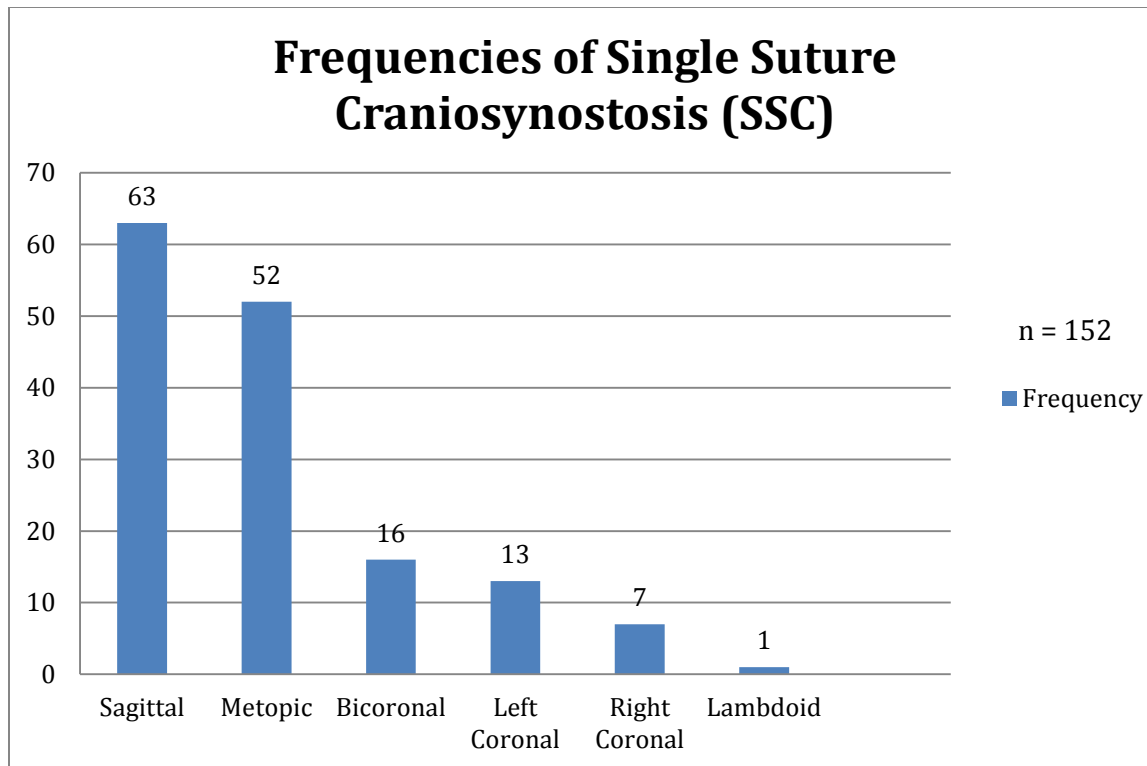


Figure 3.1. The frequencies of types of single-suture craniosynostosis.

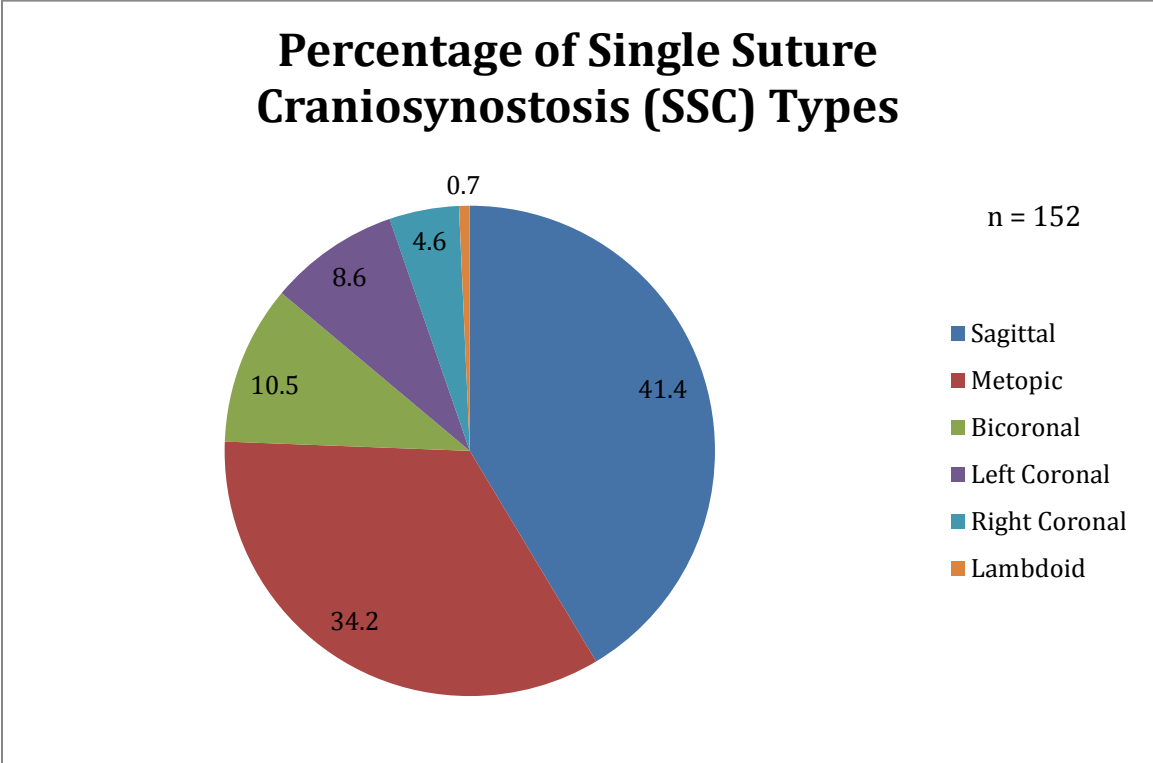


Figure 3.2. The percentage of patients with each type of single-suture craniosynostosis.



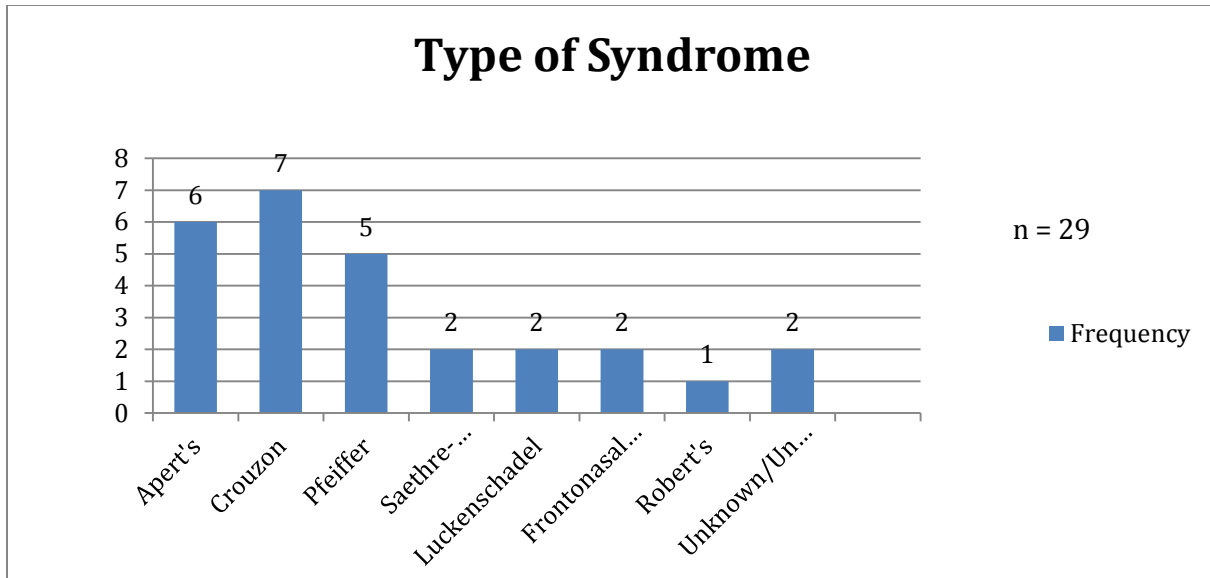


Figure 4.1. The frequencies of each type of syndromic craniosynostosis present in the sample.

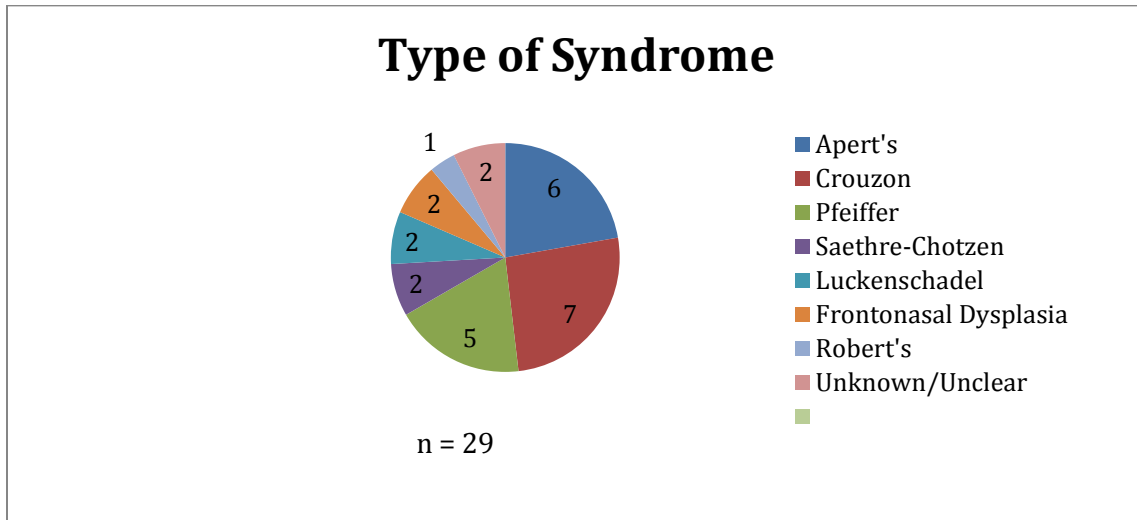
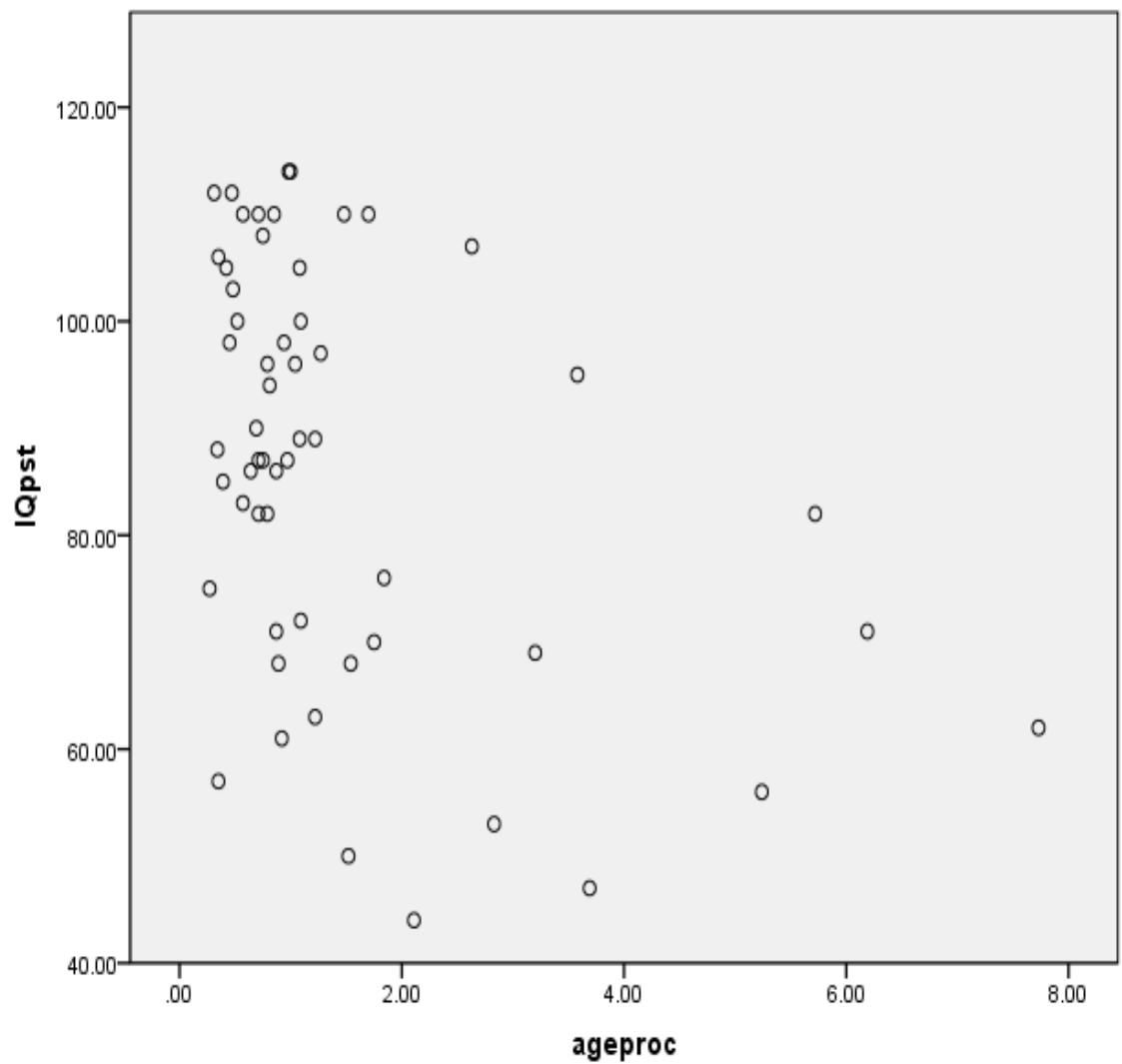


Figure 4.2. Same as Figure 4.1 showing the frequencies of each type of syndromic craniosynostosis.

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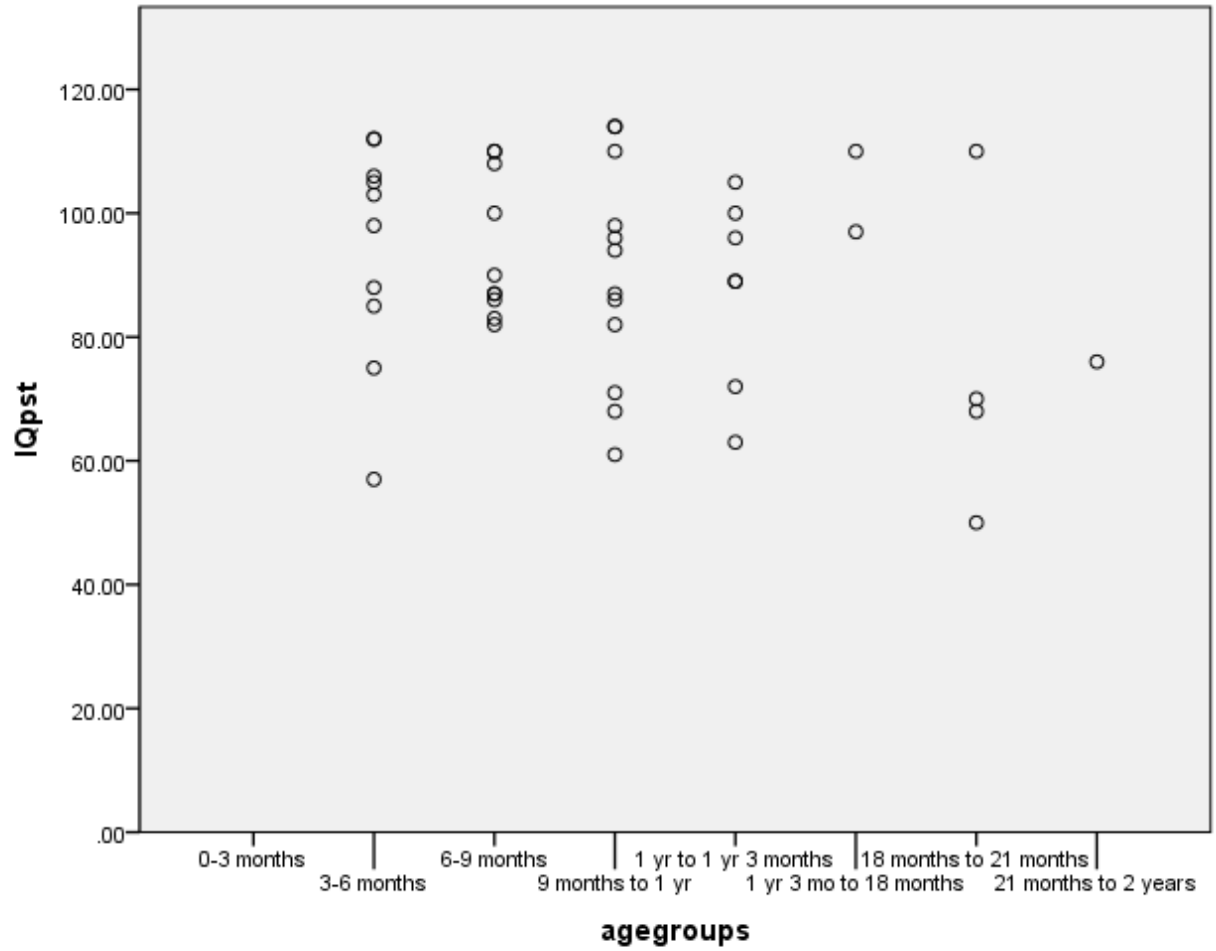
	<b>BSID</b>	<b>MSEL</b>	<b>WPPSI</b>	<b>WISC</b>	<b>BASC</b>	<b>NEPSY</b>
<b>AGE</b>	Birth – 3 yr 6 mo	Birth – 5 yr 8 mo	2 yr 6 mo – 3 yr 11 mo	6 yr – 16 yr 11 mo	2 yr – 21 yr 11 mo	3 yr – 16 yr
<b>IQ</b>			X	X		
<b>MOTOR SKILLS</b>	X	X				X
<b>LANGUAGE</b>	X	X				X
<b>INTERNALIZING SYMPTOMS</b>	X				X	
<b>BEHAVIOR</b>					X	

Table 1. Domains assessed by each neuropsychological test used for data collection in this study.



Graph 1. Pearson's  $r$  correlation of the age at which a patient received surgical intervention, and the IQ obtained from testing post-procedure.

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Graph 2. Pearson's  $r$  correlation of the age at which a patient received surgery (0-24 mo.) and IQ scores post surgery.

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	<b>Internalizing Symptoms: Presurgery</b>	<b>Internalizing Symptoms: Post-surgery</b>	<b>Language: Presurgery</b>	<b>Language: Post-surgery</b>
<i>N</i>	2	7	16	11
<i>Mean</i>	107.50	114.00	92.69	95.18
<i>SD</i>	10.61	16.11	12.85	19.68

Table 2. Sample size, mean, and standard deviation of sub-samples in patients with sagittal craniosynostosis.

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	<b>Behavior: Presurgery</b>	<b>Behavior: Post-surgery</b>	<b>Language: Presurgery</b>	<b>Language: Post-surgery</b>	<b>Gross Motor: Presurgery</b>	<b>Gross Motor: Post- surgery</b>
<i>N</i>	0	2	7	8	3	4
<i>Mean</i>	-	128.50	88.43	94.25	77.67	100.50
<i>SD</i>	-	36.06	24.81	16.28	27.30	19.88

Table 3. Sample size, mean, and standard deviation of sub-samples in patients with metopic craniosynostosis.