Review Article

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The Impact of Neurosurgery on the Neuropsychological Development of Patients with Craniosynostosis: A Scoping Review

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Introduction: Neuropsychological assessment evaluates the cognitive profile by identifying deficits and enabling monitoring. At HRAC-USP, patients with craniosynostoses undergo this assessment due to impacts on neuropsychomotor development (DNPM). These conditions may cause cranial fusion, restricting brain growth and adversely affecting cognitive functions. Evaluation is crucial for guiding therapeutic and surgical approaches and ensuring better outcomes. Objective: To identify how surgical procedures impact the neuropsychological profile of patients with syndromic and isolated craniosynostoses through a scoping review.
Methodology: This scoping review was conducted in March 2024 using PubMed, Embase, BVS/Lilacs, and Scopus databases; 1,215 articles were found and 18 original articles (E1-18) in English were selected that addressed the question: "What is the impact of surgery on neuropsychological aspects in patients with craniosynostoses?"
Results: Studies on craniosynostosis highlighted the predominance of isolated cases (83.3%), with surgeries performed before one year of age associated with better cognitive outcomes. The cranial expansion technique (38.8%) was effective in non-syndromic cases, while cranial remodeling showed varied results. After surgery, there were overall improvements in DNPM, but also risks of language delay and motor deficits, especially in patients operated on at older ages.
Conclusion: Surgery for craniosynostosis can improve neuropsychological outcomes, especially when performed early and with appropriate surgical techniques. More invasive procedures may lead to better outcomes, including increased Total Intelligence Quotient and better academic performance. It is crucial to consider such factors to optimize outcomes, reduce neurodevelopmental risks, and continuously monitor language and executive functions.

Keywords: neuropsychological tests, craniosynostoses, neuropsychology

INTRODUCTION

Neuropsychological assessment consists of cognitive evaluation, which allows qualitative and quantitative analysis of the cognitive profile - cognitive functions (praxis, executive functions, etc.) and intellectual functions of the individual, identifying early deficits and ensuring adequate follow-up [1,2].

This practice addresses demands regarding the patient's condition; therefore, cognitive evaluation assists in complementing medical exams, serving as a parameter for the cognitive functions' baseline in surgical procedures or providing support in the elaboration of therapeutic/rehabilitative approaches after the patient's discharge, aiming to benefit this process [3,4].

Given that congenital conditions affecting the skull cause adversities in neuropsychomotor development (NPMD) and the execution of certain cognitive functions [5-7], neurosurgery patients undergo this assessment at the Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo (HRAC-USP). The largest evaluated population consists of patients diagnosed with craniosynostoses, isolated or syndromic, such as Apert Syndrome, Crouzon, Pfeiffer, and Saethre-Chotzen.

Due to their syndromic nature, great variability exists in clinical and neurocognitive characteristics [8]. Within these conditions, the predominance of the fusion of more than one cranial suture is observed, restricting brain growth, and increasing intracranial pressure (ICP), negatively impacting NPMD [7,9,10].

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It is known that neuropsychological functions (cognition, executive functions, praxis, language, behavior, among others) can suffer impacts, even subtle ones, due to increased ICP. Still, these can be reduced with early surgical intervention. However, debates regarding the optimal age for intervention remain; some authors define it as between 9 and 12 months of age [8,10,11].

In this context, through a scoping review, the present study aims to identify how surgery procedures impact the neuropsychological profile of patients with syndromic and isolated craniosynostoses.

MATERIALS AND METHODS

This study constitutes a scoping review per the methodology outlined by the Joanna Briggs Institute [12], and to ensure methodological rigor, the PRISMA tool adapted for Scoping Review was employed [13] (Figure 1).

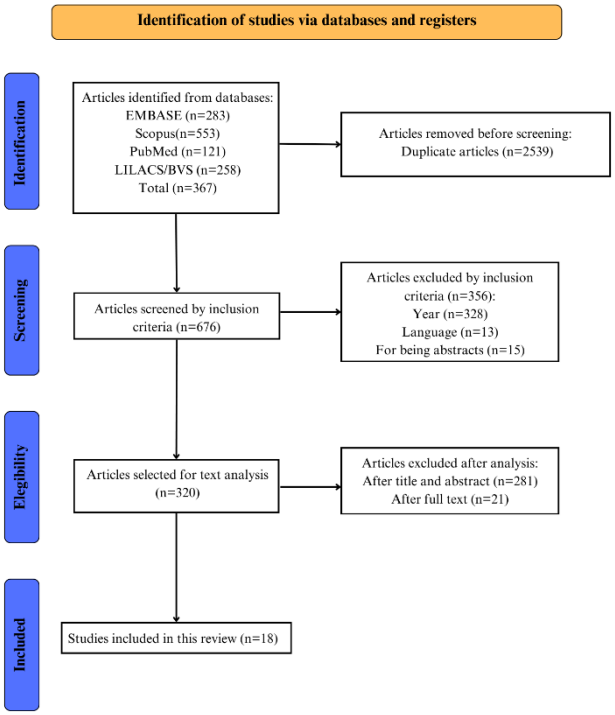

Figure 1- Flowchart of the study selection process, adapted from PRISMA-Sc

This research followed the 22 sub-steps outlined by Tricco et al. [13] to maintain scientific rigor which includes Title; Structured Abstract; Introduction - rationale and objectives -; Methods - protocol and registration, eligibility criteria, information sources, search, study selection, data collection process, data items, risk of bias in each study, summary measures, synthesis of results, risk of bias across studies, additional analyses -; Results - study selection, characteristics; critical appraisal; individual study results and synthesis of results; Discussion - summary of evidence, limitations, conclusions -; and Funding.

The Patient, Intervention, Comparison, and Outcome (PICO) [14] framework was used to devise the search strategy, as follows: P - individuals with syndromic and isolated craniosynostosis; I- surgical correction; C- individuals with typical neurodevelopment; and O- studies that testes neurodevelopmental and cognition outcomes in children after and/or before surgery. Considering this definition, the following guiding question was formulated: "What is the impact of surgery on neuropsychological aspects in patients with craniosynostosis?".

Research studies published in English or Portuguese after 2015 were eligible for inclusion, provided they contained information regarding surgery and developmental assessment. Review articles were excluded, as studies do not address the research question, posters from conference proceedings, expert opinions, and brochures.

The articles were researched with librarian assistance from March 05th to 7th, 2024 using the following databases: National Library of Medicine (PubMed/MEDLINE), Embase, Scopus, and Latin American and Caribbean Health Sciences Literature (LILACS/BVS).

Health descriptors (DeCS/MESH), keywords, and their alternative terms were used for the search. Boolean operators "or" and "and" were employed, and the following terms were defined: (Acrocephalosyndactylia OR Craniofacial Dysostosis OR Craniosynostoses) AND (Neuropsychological Test OR Cognitive Tests OR Neuropsychology OR Neurosurgery).

The search yielded 1215 articles via databases. After removing 539 duplicates with Mendeley software, 676 articles remained for screening. Through the Rayyan - Intelligent Systematic Review tool, 354 studies were excluded for failing to meet the inclusion criteria. After screening titles and abstracts, 281 articles were deleted, leaving 41 for full-text screening. Ultimately, eighteen studies met the criteria for inclusion (Figure 1).

The articles numbered 01 to 18 were called "study" for analysis. The results were presented in the form of tables and discursive reports.

RESULTS

Of the 18 studies included in the analysis, all were written in English. Over the last decade, 8 (44.4%) were published in the United States, followed by South Korea with 3 (14.2%) publications on the subject, and with one publication (4.7%) in Brazil, United Kingdom, Mexico, Turkey, France, Netherlands, and India (table 1).

Table 1 - Study selection according to author, year, type, and country of publication.

	Authorship	Year of publication	Title	Type of study	Country where the study was carried out
S1	Collet, B. et al.	2017	Attention and executive function in children with and without single-suture craniosynostosis	Observational	United States
S2	Moreno-Villagómez, J. et al.	2023	Cognitive performance in preschoolers with non-syndromic craniosynostosis undergoing surgery: A comparison with typically developing children	Comparative	Mexico
S3	Wu, R. et al.	2020	Comparison of Neurocognitive Outcomes in Postoperative Adolescents with Unilateral Coronal Synostosis	Prospective cohort	United States
S4	Lee, M. et al.	2017	Correction of Sagittal Craniosynostosis Using Distraction Osteogenesis Based on Strategic Categorization	Prospective Observational	South Korea
S5	Aksoğan, Y. et al.	2024	Evaluation of neurocognitive and social developments after craniosynostosis surgery	Case control	Turkey
S6	Lee, M. et al.	2015	Expansion and compression distraction osteogenesis based on volumetric and neurodevelopmental analysis in sagittal craniosynostosis	Prospective cohort	South Korea
S7	Kapp-Simon, K. et al.	2016	Language, learning, and memory in children with and without single-suture craniosynostosis	Case control	South Korea
S8	Chuang, C. et al.	2021	Long-Term Neurocognitive Outcomes in Sagittal Synostosis: The Impact of Reoperation.	Retrospective study	United States
S9	Alperovich, M. et al.	2021	Long-Term Neurocognitive Outcomes of Spring-Assisted Surgery versus Cranial Vault Remodeling for Sagittal Synostosis	Retrospective cohort	United States
S10	Fontana, S. et al.	2018	Longitudinal Assessment of Developmental Outcomes in Infants Undergoing Late Craniosynostosis Repair.	Non-randomized prospective	United States
S11	Lynn, J. et al.	2023	Longitudinal Assessment of Neurodevelopment in Patients With Nonsyndromic Single-Suture Craniosynostosis: A Retrospective Review of 66 Patients	Retrospective review	United States
S12	Verlut, I. et al.	2019	Neuropsychological consequences of craniosynostosis: Non-syndromic scaphocephaly	Observational cohort	France
S13	Den Otterlander, B. et al.	2020	Saethre–Chotzen syndrome: long-term outcome of a syndrome-specific management protocol	Retrospective cohort	Netherlands
S14	Cradock, M. et al.	2015	Sex differences in the neurodevelopment of school-age children with and without single-suture craniosynostosis	Longitudinal	United States
S15	Lynn, J. et al.	2023	Surgical Timing and Neurocognitive Development among Patients with Craniosynostosis: Analysis of Confounders	Retrospective cohort	United States
S16	Maximino, L. et al.	2017	Syndromic craniosynostosis: neuropsycholinguistic abilities and imaging analysis of the central nervous system	Cross-sectional	Brazil
S17	Uppar, A. et al.	2022	Syndromic craniosynostosis: Objective and parent-reported outcome measurements after cranio-facial remodelling surgeries	Retrospective cohort	India
S18	Edwards-Bailey, L. et al.	2022	The Craniofacial Collaboration UK: Developmental Outcomes in 5-Year-Old Children With Metopic Synostosis	Longitudinal prospective	United Kingdom

Table 2 categorizes the studies based on the neuropsychological assessment methodology used and the functions evaluated in the population. The predominant cognitive assessment methodology applied across the 18 studies reviewed was the Wechsler Intelligence Scale and its various iterations, utilized in nine studies (50%) to evaluate the intelligence quotient (IQ) [6,15-22]. Afterward, the Bayley Scales of Infant and Toddler Development were employed in six studies (33.3%) to evaluate NPMD [23-28], and the Behavior Rating Inventory of Executive Function (BRIEF) was utilized in four articles (22.2%) aiming to appraise executive functions [15-17,21].

However, in Verlut et al. [21], two tools were applied to evaluate emotions and anxiety: the Positive and Negative Affect Schedule and the Revised Children's Manifest Anxiety Scale. One study did not specify the instruments used in the assessment [29].

Table 3 presents the studies according to diagnosis, surgical procedures, ages, and outcomes.

Only three studies (16.6%) included patients diagnosed with syndromic craniosynostoses, such as Apert syndrome [6,30], Crouzon syndrome [6,30], Pfeiffer Syndrome [30], and Saethre-Chotzen syndrome [29,30]. Among the 15 (83.3%) studies that investigated isolated craniosynostosis, sagittal (12; 29.7%) [16-24,26-28], and metopic (10; 27%) [18-26,32] craniosynostosis were the most studied type, followed by unicoronal synostosis (8; 21.6%) [15,18-20,22-23,25-26], Lambdoid (5; 13.5%) [18-20,22,24], bicoronal (2; 5.4%) [24,26], and multisuture (1; 2.7%) [20] craniosynostosis were less frequently represented.

Considering the type of surgical procedure, a few studies associated more than one surgical technique[16,23,25-26,30], nevertheless the most common technique used was cranial vault expansion (CVE) related in seven (38.8%) studies [16-19,23,25,29], followed by six (33.3%) articles that mentioned cranial vault remodeling (CVR) [15,20,22,24,26,30].

Other three techniques were mentioned in other studies, such as fronto-orbital advancement (4; 22.2%) [23,25-26,30] distraction osteogenesis (3; 16.6%) [16,27-28], and Le Fort III osteotomy (1; 5.5%) [30]. Another three (16.6%) did not specify the surgical technique used [6,21,31].

Regarding the age during surgery, nine studies (50%) involved populations operated on when they were less than or equal to 1 year old [15-18,20-21,24,26,29]. In comparison, four studies (22.2%) involved patients who underwent surgery after turning 1 year old [19,22,23,25]. Three studies (16.7%) showed high variability in age, ranging from 3 months to 8 years old [27-28,30]. Two studies (11.1%) did not provide information about the age at the time of procedure [6,31].

As stated in table 2, the most evaluated functions were IQ, NPMD, and executive functions. Notably, the results described below consider the assessment outcomes after the surgery.

Of the nine studies (50%) that assessed the Full-Scale Intelligence Quotient (FSIQ) [6,15-22], only two (22.2%) [18-19] did not present results regarding the FSIQ, separately investigating verbal and memory functions. Meanwhile, of the remaining seven studies, three (33.3%) reported below-average FSIQ [6,16,22], while the remaining four (44.4%) studies indicated FSIQ within the expected range for the age group [15,17,20-21].

In the six studies (33.3%) [23-28], employing the Bayley Scales of Infant and Toddler Development [32] to assess NPMD, improvement was observed across all sections of the tool: "Personal Social Development," "Fine Motor Development," "Gross Motor Development," and "Language Cognitive Development." Despite the improvement all authors described nonsignificant changes before and after surgery, also some authors noted a potential risk of language and motor delay compared to typical subjects, even after surgery [23-24,26].

Among the four studies (22.2%) [15,17-18,21], that assessed executive functions using the BRIEF instrument, three (75%) indicated deficits in the evaluated function even after surgery [17-18,21].

DISCUSSION

When identifying the type of craniosynostosis, 83.3% of the studies focused on isolated craniosynostosis [15-28,31], this prevalence may be attributed to its higher occurrence in the population when compared to syndromic cases, with a prevalence rate of one in 2,000 to 2,500 live births [33].

While debates persist regarding the optimal age for surgery, there is a consensus that it is most beneficial to perform the procedure before the child reaches one year of age [8,10,11,34]. This can be observed in the results among the studies assessing FSIQ (50%) [6,15-22], where 33.3% reported below-average FSIQ, particularly in patients operated on later in childhood [6,16,22]. It is noteworthy that two of the studies [6] focused solely on patients with syndromic craniosynostosis, a group known to face heightened neurodevelopmental risks due to additional complications associated with their syndromes [8-10].

Significantly, the remaining 44.4% of studies indicated that the FSIQ fell within the expected range for the respective age group, with all patients having undergone surgery before the age of one year [15,17,20-21].

Table 2 - Neuropsychological assessment methodology used and the functions evaluated in the population.

Assessment method	Functions evaluated	Studies
A Developmental Neuropsychological Assessment (NEPSY-II)	Attention and Executive Functioning; Language; Memory and Learning; Sensorimotor; Social Perception; and Visuospatial Processing	18, 19, 21
Achenbach System of Empirically Based Assessment (ASEBA)	Adaptative behavior	18
Ages and Stage Questionnaire (ASQ-3)	Emotions and Behavior	31
Bayley Scales of Infant and Toddler Development, 2nd Edition (Bayley-II)	Neurodevelopment	27, 28
Bayley Scales of Infant and Toddler Development, 3rd Edition (Bayley-III)	Neurodevelopment	23, 24, 25, 26
Beery-Buktenica Developmental Test of Visual-Motor Integration (BEERY™ VMI)	Visual-motor skills	15, 16, 17
Behavior Assessment System of Children, 2nd Edition (BASC-2)	Emotions and Behavior	17
Behavior Rating Inventory of Executive Function (BRIEF)	Executive functions	15, 17, 18,21
Brunet–Lézine Scale of Development Revised (BLR)	Neurodevelopment	21
Child Behavior Checklist (CBCL)	Internalizing and externalizing behaviors	15
Children's Communication Checklist-2 (CCC-2)	Pragmatic language	19
Children's Memory Scale	Memory and attention	19
Comprehensive Test of Phonological Processing (CTOPP-2)	Phonological skills	20
Parent-reported outcome measurement (PROM)	Physical/functional and social well-being; Scholastic performance, and Intelligence	30
Peabody Picture Vocabulary Test (PPVT-IV)	Receptive language	6
Scale of Motor Functional Development (DF-MOT)	Motor skills	21
School Achievement Test	Academic skills (reading, writing, and arithmetic)	6
Strengths and Difficulties Questionnaire (SDQ)	Emotions and Behavior	31
Test of Everyday Attention in Children (TEA-Ch)	Attention	18
Test of Word Reading Efficiency (TOWRE)	Reading skills	20
The Wide Range Assessment Test, 4th Edition (WRAT-4)	Academic skills (reading, arithmetic, comprehension, and spelling)	20
Token Test	Verbal comprehension, receptive language, working memory, inhibitory control, and attention	6, 19
Wechsler Abbreviated Scale of Intelligence (WASI)	Intelligence	6, 15, 16, 17
Wechsler Fundamentals: Academic Skills (WF)	Academic skills (reading, arithmetic, comprehension, and spelling)	15, 16, 17
Wechsler Intelligence Scale for Children, 3rd Edition (WISC-III)	Intelligence	6
Wechsler Intelligence Scale for Children, 4th Edition (WISC-IV)	Intelligence	18, 19, 20
Wechsler Intelligence Scale for Children, 5th Edition (WISC-IV)	Intelligence	21
Wechsler Intelligence Scale for Children, Revised Edition (WISC-R)	Intelligence	21
Wechsler Preschool and Primary Scale of Intelligence- 3rd and 4th Edition (WPPSI-III; WPPSI-IV)	Intelligence	21, 22

Table 3 - Population, Surgical procedure, age, and outcomes from the studies selected.

	Article	Population	Diagnosis		Type/Suture	Assessment age	Surgical procedure	Time of the procedure	Results
			Isolated	Syndromic					
S1	Collet B., et al (2017)	179	X		Lambdoid; Metopic; Unicoronal; Sagittal	18 mos; 36 mos; and 7 yo	Cranial vault expansion	Between 5 mos to greater than 15 mos	High impairment in attention and executive functions, particularly in inhibitory control.
S2	Moreno-Villagómez, J. et al. (2023)	31	X		Lambdoid; Metopic; Unicoronal; Sagittal; Multisuture	3 to 5yo and 11 mos	Cranial vault remodeling	13 mos	Lower performance on Verbal IQ and Total IQ; Children with unilateral coronal craniosynostosis performed worse than the others.
S3	Wu, R. et al. (2020)	20	X		Unicoronal	12 yo (range age)	Cranial vault remodeling	8 mos (range age)	Verbal and executive intelligence, along with academic reading and writing skills, fell within the expected range for the respective age group. Mathematical and motor skills were below average. Children operated on before 7 mos had higher motor coordination scores, while strabismus was more common in patients operated on at an older age.
S4	Lee, M. et al. (2017)	25	X		Sagittal	5 months to 7yo (2mo before and 1yo after the surgery)	Distraction ostegenesis	5 mos to 8 yo	Significant improvement in cognitive and psychomotor development after surgery was observed after the operation when performed before 1 yo.
S5	Aksoğan, Y. et al. (2024)	73	X		Metopic; Unicoronal; Sagittal; Bicoronal	2 to 6 yo	Cranial vault remodeling; Fronto-orbital advancement	3 mos to 1 yo	Substantial risk of delay in language and fine motor skills after surgery.
S6	Lee, M. et al. (2015)	16	X		Sagittal	5 mos to 7 yo (2 mos before and 1 yo after the surgery)	Distraction ostegenesis	1 to 8 yo	Significant improvement in mental and psychomotor development.
S7	Kapp-Simon, K. et al. (2016)	179	X		Lambdoid; Metopic; Unicoronal; Sagittal	7 yo	Cranial vault expansion	1 to 2 yo	Memory and language abilities were impacted.

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S8	Chuang, C. et al. (2021)	47	X		Sagittal	5 to 16 yo	Cranial vault expansion	6 mos and reoperation between 5 to 16 yo	Patients who underwent reoperation showed better IQ, academic achievement, and visuomotor integration performance than those who did not undergo reoperation and whose surgery occurred later.
S9	Alperovich, M. et al. (2021)	75	X		Sagittal	10 yo (range age)	Cranial vault expansion; Distraction osteogenesis spring-assisted	4.8 mos (range age)	Patients operated on with the cranial vault expansion technique achieved better outcomes in intelligence and visuomotor function compared to those operated on with distraction osteogenesis using expanding springs.
S10	Fontana, S. et al. (2018)	5	X		Metopic; Unicoronal	1 to 3yo 7mo	Cranial vault expansion; Fronto-orbital advancement	13 to 43 mos	Overall improvement in all cases with developmental delay. A baby who did not exhibit any delay remained within the average range after surgery.
S11	Lynn, J. et al. (2023)	66	X		Sagittal; Metopic; Unicoronal	1 to 3yo 6mo	Cranial vault expansion; Fronto-orbital advancement	3 mos to 1 yo 6 mos	Patients exhibited delays compared to their age group, and no significant changes were observed after surgery.
S12	Verlut, I. et al. (2019)	29	X		Sagittal; Metopic	5 to 14 yo (scaphocephaly); 6 to 11 yo (trigonocephaly)	Nonspecified	Between less than and greater than 6 mos (scaphocephaly); less than or equal 8 mos (trigonocephaly)	Children with scaphocephaly had IQ scores within the expected range for their age but significant deficits in executive and attentional functions, while cases with trigonocephaly showed deficits across all areas.
S13	Den Otterlander, B. et al. (2020)	30		X	Saethre-Chotzen Syndrome	range age 13 yo	Cranial vault expansion	9.6 mos (range age)	Three children attended a specialized school with an individualized curriculum, primarily due to intellectual disability, with one child having mild intellectual disability. Additionally, twelve children had language delays and three patients were enrolled in nursery school.
S14	Cradock, M. et al. (2015)	180	X		Metopic; Sagittal; Unicoronal; Lambdoid	7 yo	Cranial vault remodeling	Range age 9.1 mos (less or equal and great than 6 mos)	Males consistently scored lower than females across all outcomes, indicating a higher likelihood of learning problems among males compared to females.

S15	Lynn, J. et al. (2023)	227 (157 early / 70 late)	X		Bicoronal; Sagittal; Metopic; Lambdoid	7.3 mos (early) and 13.8 mos (late) (range age)	Cranial vault remodeling	11.2 mos (range age)	The late operation group showed significant language and cognitive delay. There was no difference in motor performance.
S16	Maximino, L. et al. (2017)	18		X	Apert Syndrome; Crouzon Syndrome	6 to 31 yo	Nonspecified	Uninformed	8 individuals exhibited learning disorders, 3 had language impairments, and 4 displayed intellectual disabilities, while 9 had average IQ scores. It was noted that learning disorders were linked to mild cognitive impairments
S17	Edwards-Bailey, L. et al. (2022)	91	X		Metopic	4 yo 6 mos to 5 yo to 5 mos	Nonspecified	Uninformed	All individuals exhibited deficits in communication, gross and fine motor skills, personal-social interactions, and emotional regulation. Additionally, hyperactivity was observed among the group.
S18	Uppar, A. et al. (2022)	15		X	Apert Syndrome; Crouzon Syndrome; Saethre-Chotzen Syndrome; and Pfeiffer Syndrome	1 to 7 yo 6 mos	Cranial vault remodeling with fronto-orbital advancement; Le Fort III osteotomy	6 mos to 7 yo	IQ and academic skills were within the expected range for their age group.

*mos = months of age; *yo= years of age.

One study [17] underscores the enhancement in FSIQ, academic performance, and visuomotor integration among patients who underwent reoperation between the ages of 5 to 16 years, after the initial surgery performed before the age of one.

Conversely, studies using the Bayley Scales of Infant and Toddler Development [32] for neurodevelopmental assessment observed improvement across various domains post-surgery, albeit with a potential language and motor delay risk compared to typical subjects [24,26-28]. Three of the selected studies that identified delays involved a mixed population, with some individuals having undergone surgery after one year old [23-24,28].

Additionally, studies assessing executive functions using the BRIEF instrument noted deficits persisting after surgery, particularly in patients operated on at older ages [10-17-18]. These findings underscore the importance of considering age in surgical decision-making and postoperative neurocognitive monitoring to optimize outcomes.

Some authors recognize age at surgical time as a significant factor in pursuing better outcomes because, during this period of development the skull bones are still flexible enough to undergo remodeling more easily, reducing the impacts of increased ICP and craniofacial dysmorphisms related to brain growth [8,35]. Also, Bruce et al. [36] highlight syndromic cases with elevated perioperative risk due to supplementary factors, which can be reduced in early-age surgery.

The correlation between age and surgical procedure in managing craniosynostosis involves multiple facets. Among the studies, CVE was the most common technique (38.8%), associated with improved neurodevelopmental outcomes in some cases, all populations included were not syndromic. Regarding age, three of five studies involved infants operated on within the first year of life [18,23,25], while a

substantial portion included patients operated on after age one.

The CVE technique alleviates increased intracranial pressure by allowing skull expansion and facilitating brain growth [35,37]. Studies indicate that early whole vault cranioplasty, performed before 6 months of age, yields superior intelligence quotient and achievement scores compared to minor invasive surgery, such as strip craniectomy [38-41].

Unlike that, cranial vault remodeling (33.3%) [15,20,22,24,26,30] yielded mixed results regarding neurocognitive improvement. Five studies [15,20,22,24,26] described lower outcomes after the surgery; three of these studies [15,22,24] presented patients operated on after one year old.

The limited space for a developing brain and elevated intracranial pressure result in neuroanatomical changes extending beyond the areas directly affected by the fused suture [19]. This phenomenon is particularly notable among individuals with isolated craniosynostosis, who often experience persistent neuropsychological deficits that become more pronounced during school age [17-18,20,22].

Only one study [30] focused on individuals with syndromic craniosynostosis who underwent multiple surgeries using various techniques throughout their lifetime, including CVR with fronto-orbital advancement and Le Fort III osteotomy. Remarkably, all patients demonstrated FSIQ and academic skills within age-appropriate ranges. This positive outcome could be attributed to combining CVR with fronto-orbital advancement, which aims to reshape the skull and create additional space for brain growth [40].

CONCLUSION

Surgical intervention for craniosynostosis can positively impact neuropsychological outcomes, particularly when performed early in life and with appropriate selection techniques. Operative treatment performed before age one, combined with the selection of a proper surgical technique, is essential. More invasive procedures may offer better outcomes, enhancing neuropsychological results, including elevated FSIQ scores and enhanced academic performance. Overall, surgery for craniosynostosis management requires careful consideration of various factors to optimize results and minimize neurodevelopmental risks.

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none

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Considering the present study is a review, there is no need for Ethic Committe evaluation.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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Artificial intelligence

No artificial intelligence was not used in this paper work

CONTRIBUTIONS

-**Jéssica Luchi Ferreira**: Formal Analysis, Investigation, Methodology, Writing – original draft, Writing – review & editing
-**Igor José Nogueira Gualberto**: Data curation, Methodology, Writing – review & editing
-**Mariani da Costa Ribas**: Conceptualization, Supervision, Writing – review & editing
-**Michele Madeira Brandão:** Supervision
-**Cristiano Tonello**: Conceptualization, Supervision

REFERENCES

1. Winograd, M., Martins de Jesus, M. V., & Uehara, E. The qualitative aspects in practice of the neuropsychology assessment. Ciências & Cognição. 2012;17(2), 02-13.
2. Lezak, M. D. Neuropsychological assessment. Oxford University Press, USA; 2004.
3. Lazzaretti, CT. Manual de Psicologia Hospitalar: Coletânea ConexãoPsi. 21. ed. Curitiba: [s.n.]; 2007.
4. Mäder, B. Caderno de Psicologia Hospitalar: considerações sobre assistência, ensino, pesquisa e gestão. Curitiba: CRP-PR, 2016.
5. Kutkowska-Kaźmierczak A, Gos M, Obersztyn E. Craniosynostosis as a clinical and diagnostic problem: molecular pathology and genetic counseling. J Appl Genet. 2018 May;59(2):133-147. doi: 10.1007/s13353-017-0423-4. Epub 2018 Feb 1. Erratum in: J Appl Genet. 2018 May;59(2):149-150. doi: 10.1007/s13353-018-0438-5. PMID: 29392564.
6. Maximino LP, Ducati LG, Abramides DVM, Corrêa CC, Garcia PF, Fernandes AY. Syndromic craniosynostosis: neuropsycholinguistic abilities and imaging analysis of the central nervous system. Arq Neuropsiquiatr. 2017 Dec;75(12):862-868. doi: 10.1590/0004-282X20170171. PMID: 29236889.

7. Brandao MM, Tonello C, Parizotto I, Machado LB, Alonso N. Analysis of intracranial pressure waveform using a non-invasive method in individuals with craniosynostosis. Childs Nerv Syst. 2024 Jan;40(1):145-152. doi: 10.1007/s00381-023-06092-y. Epub 2023 Jul 26. PMID: 37493721.
8. Sawh-Martinez R, Steinbacher DM. Syndromic Craniosynostosis. Clin Plast Surg. 2019 Apr;46(2):141-155. doi: 10.1016/j.cps.2018.11.009. PMID: 30851747.
9. Brooks ED, Beckett JS, Yang J, Timberlake AT, Sun AH, Chuang C, Persing JA. The Etiology of Neuronal Development in Craniosynostosis: A Working Hypothesis. J Craniofac Surg. 2018 Jan;29(1):49-55. doi: 10.1097/SCS.0000000000004040. PMID: 29049144.
10. van der Vlugt JJB, van der Meulen JJMN, van den Braak RRJC, Vermeij-Keers C, Horstman EGC, Hovius SER, Verhulst FC, Wierdsma AI, Lequin MH, Okkerse JME. Insight into the Pathophysiologic Mechanisms behind Cognitive Dysfunction in Trigonocephaly. Plast Reconstr Surg. 2017 Apr;139(4):954e-964e. doi: 10.1097/PRS.0000000000003179. PMID: 28350677.
11. Ecklund-Johnson E, Pearson CM. Foundations of contemporary neuropsychology. In: Neurosurgical Neuropsychology: The Practical Application of Neuropsychology in the Neurosurgical Practice. Elsevier; 2018. p. 27–37.
12. Peters MDJ, Marnie C, Tricco AC, Pollock D, Munn Z, Alexander L, McInerney P, Godfrey CM, Khalil H. Updated methodological guidance for the conduct of scoping reviews. JBI Evid Synth. 2020 Oct;18(10):2119-2126. doi: 10.11124/JBIES-20-00167. PMID: 33038124.
13. Tricco AC, Lillie E, Zarin W, O'Brien KK, Colquhoun H, Levac D, Moher D, Peters MDJ, Horsley T, Weeks L, Hempel S, Akl EA, Chang C, McGowan J, Stewart L, Hartling L, Aldcroft A, Wilson MG, Garritty C, Lewin S, Godfrey CM, Macdonald MT, Langlois EV, Soares-Weiser K, Moriarty J, Clifford T, Tunçalp Ö, Straus SE. PRISMA Extension for Scoping Reviews (PRISMA-ScR): Checklist and Explanation. Ann Intern Med. 2018 Oct 2;169(7):467-473. doi: 10.7326/M18-0850. Epub 2018 Sep 4. PMID: 30178033.
14. Higgins JPT, Green S (eds). Cochrane Handbook for Systematic Reviews of Interventions, Version 5.1.0. London: The Cochrane Collaboration; 2011.
15. Wu RT, Gabrick KS, Singh A, Landi N, Taylor JA, Bartlett SP, Persing JA, Alperovich M. Comparison of Neurocognitive Outcomes in Postoperative Adolescents with Unilateral Coronal Synostosis. Plast Reconstr Surg. 2020 Sep;146(3):614-619. doi: 10.1097/PRS.0000000000007067. PMID: 32842112.
16. Alperovich M, Runyan CM, Gabrick KS, Wu RT, Morgan C, Park SE, Chapman LA, Couture DE, David LR, Persing JA. Long-Term Neurocognitive Outcomes of Spring-Assisted Surgery versus Cranial Vault Remodeling for Sagittal Synostosis. Plast Reconstr Surg. 2021 Mar 1;147(3):661-671. doi: 10.1097/PRS.0000000000007640. PMID: 33620934.
17. Chuang C, Chaunzwa TL, Wu R, Singh A, Patel A, Yang JF, Hashim PW, Travieso R, Terner JS, Mayes LC, Duncan CC, Jane JA Jr, Lin KY, Bridgett DJ, Persing JA. Long-Term Neurocognitive Outcomes in Sagittal Synostosis: The Impact of Reoperation. J Craniofac Surg. 2021 Jan-Feb 01;32(1):58-61. doi: 10.1097/SCS.0000000000006909. PMID: 33394632.
18. Collett BR, Kapp-Simon KA, Wallace E, Cradock MM, Buono L, Speltz ML. Attention and executive function in children with and without single-suture craniosynostosis. Child Neuropsychol. 2017 Jan;23(1):83-98. doi: 10.1080/09297049.2015.1085005. Epub 2015 Sep 18. PMID: 26381123; PMCID: PMC5181850.
19. Kapp-Simon KA, Wallace E, Collett BR, Cradock MM, Crerand CE, Speltz ML. Language, learning, and memory in children with and without single-suture craniosynostosis. J Neurosurg Pediatr. 2016 May;17(5):578-88. doi: 10.3171/2015.9.PEDS15238. Epub 2016 Jan 1. PMID: 26722698.
20. Cradock MM, Gray KE, Kapp-Simon KA, Collett BR, Buono LA, Speltz ML. Sex differences in the neurodevelopment of school-age children with and without single-suture craniosynostosis. Childs Nerv Syst. 2015 Jul;31(7):1103-11. doi: 10.1007/s00381-015-2671-0. Epub 2015 Apr 22. PMID: 25899849.
21. Verlut I, Mottolese C, Szathmari A, Banvillet E, Poncet A, Blache A, Rousselle C, Gleizal A, Paulus C, Di Rocco F. Le retentissement neuropsychologique de la cranio sténose: cas de la scaphocéphalie non syndromique [Neuropsychological consequences of craniosynostosis: Non-syndromic scaphocephaly]. Neurochirurgie. 2019 Nov;65(5):330-336. French. doi: 10.1016/j.neuchi.2019.09.018. Epub 2019 Oct 1. PMID: 31585152.
22. Moreno-Villagómez J, Yáñez-Téllez G, Prieto-Corona B, Seubert-Ravelo AN, García A, Hernández-Echeagaray E. Cognitive performance in preschoolers with non-syndromic craniosynostosis undergoing surgery: A comparison with typically developing children. Appl Neuropsychol Child. 2023 Feb 25:1-7. doi: 10.1080/21622965.2023.2183476. Epub ahead of print. PMID: 36840597.
23. Lynn JV, Buchman LK, Breuler CJ, Buchman SR. Longitudinal Assessment of Neurodevelopment in Patients With Nonsyndromic Single-Suture

Craniosynostosis: A Retrospective Review of 66 Patients. J Craniofac Surg. 2023 May 1;34(3):931-935. doi: 10.1097/SCS.0000000000009140. Epub 2022 Nov 25. PMID: 36730211.

24. Lynn JV, Buchman LK, Breuler CJ, Buchman SR. Surgical Timing and Neurocognitive Development among Patients with Craniosynostosis: Analysis of Confounders. Plast Reconstr Surg. 2023 Apr 1;151(4):821-829. doi: 10.1097/PRS.0000000000010004. Epub 2023 Mar 29. PMID: 36728531.
25. Fontana SC, Belinger S, Daniels D, Tuttle M, Camarata PJ, Andrews BT. Longitudinal Assessment of Developmental Outcomes in Infants Undergoing Late Craniosynostosis Repair. J Craniofac Surg. 2018 Jan;29(1):25-28. doi: 10.1097/SCS.0000000000004024. PMID: 29040147.
26. Aksoğan Y, Kuzucu P, Soysal Acar AŞ, Şahin MB, Gücüyener K, Börcek AÖ. Evaluation of neurocognitive and social developments after craniosynostosis surgery. Childs Nerv Syst. 2024 May;40(5):1489-1499. doi: 10.1007/s00381-024-06303-0. Epub 2024 Jan 31. PMID: 38294493.
27. Lee MC, Shim KW, Park EK, Yun IS, Kim DS, Kim YO. Expansion and compression distraction osteogenesis based on volumetric and neurodevelopmental analysis in sagittal craniosynostosis. Childs Nerv Syst. 2015 Nov;31(11):2081-9. doi: 10.1007/s00381-015-2843-y. Epub 2015 Aug 1. PMID: 26231567.
28. Lee MC, Shim KW, Yun IS, Park EK, Kim YO. Correction of Sagittal Craniosynostosis Using Distraction Osteogenesis Based on Strategic Categorization. Plast Reconstr Surg. 2017 Jan;139(1):157-169. doi: 10.1097/PRS.0000000000002899. PMID: 28027243.
29. Den Ottelander BK, Van Veelen MC, De Goederen R, Van De Beeten SD, Dremmen MH, Loudon SE, Versnel SL, Van Den Ouweland AM, Van Dooren MF, Joosten KF, Mathijssen IM. Saethre-Chotzen syndrome: long-term outcome of a syndrome-specific management protocol. Dev Med Child Neurol. 2021 Jan;63(1):104-110. doi: 10.1111/dmcn.14670. Epub 2020 Sep 9. PMID: 32909287; PMCID: PMC7754116.
30. Uppar AM, Shukla D, Nayak N, Rao G, Dwarakanath S. Syndromic Craniosynostosis: Objective and Parent-Reported Outcome Measurements after Cranio-Facial Remodelling Surgeries. Pediatr Neurosurg. 2022;57(1):17-27. doi: 10.1159/000518393. Epub 2021 Nov 8. PMID: 34818259.
31. Edwards-Bailey L, Piggott K, Dalton L, Horton J, Parks C, Wright G, Kearney A. The Craniofacial Collaboration UK: Developmental Outcomes in 5-Year-Old Children With Metopic Synostosis. J Craniofac Surg. 2023 May 1;34(3):855-859. doi: 10.1097/SCS.0000000000009095. Epub 2022 Nov 4. PMID: 36329005.
32. Spencer-Smith MM, Spittle AJ, Lee KJ, Doyle LW, Anderson PJ. Bayley-III Cognitive and Language Scales in Preterm Children. Pediatrics. 2015 May;135(5). doi: 10.1542/peds.2014-3039. Epub 2015 Apr 20. PMID: 25896835.
33. Slater BJ, Lenton KA, Kwan MD, Gupta DM, Wan DC, Longaker MT. Cranial sutures: a brief review. Plast Reconstr Surg. 2008 Apr;121(4):170e-178e. doi: 10.1097/01.prs.0000304441.99483.97. PMID: 18349596.
34. Pearson GD, Havlik RJ, Eppley B, Nykiel M, Sadove AM. Craniosynostosis: a single institution's outcome assessment from surgical reconstruction. J Craniofac Surg. 2008 Jan;19(1):65-71. doi: 10.1097/SCS.0b013e31815c8aae. PMID: 18216667.
35. Shakir S, Birgfeld CB. Syndromic Craniosynostosis: Cranial Vault Expansion in Infancy. Oral Maxillofac Surg Clin North Am. 2022 Aug;34(3):443-458. doi: 10.1016/j.coms.2022.01.006. Epub 2022 Jul 2. PMID: 35787825.
36. Bruce WJ, Chang V, Joyce CJ, Cobb AN, Maduekwe UI, Patel PA. Age at Time of Craniosynostosis Repair Predicts Increased Complication Rate. Cleft Palate Craniofac J. 2018 May;55(5):649-654. doi: 10.1177/1055665617725215. Epub 2017 Dec 14. PMID: 29665342; PMCID: PMC5907921.
37. Cohen MM Jr, Kreiborg S. Birth prevalence studies of the Crouzon syndrome: comparison of direct and indirect methods. Clin Genet. 1992 Jan;41(1):12-5. doi: 10.1111/j.1399-0004.1992.tb03620.x. PMID: 1633640.
38. Chandler L, Allam O, Park KE, Smetona J, Gabrick KS, Wu RT, Morgan C, Park SE, Chapman LA, Couture DE, David LR, Runyan CM, Persing JA, Alperovich M. Spring-Assisted Strip Craniectomy Versus Cranial Vault Remodeling: Long-Term Psychological, Behavioral, and Executive Function Outcomes. J Craniofac Surg. 2020 Oct;31(7):2101-2105. doi: 10.1097/SCS.0000000000006806. PMID: 32890160.
39. Hashim PW, Patel A, Yang JF, Travieso R, Terner J, Losee JE, Pollack I, Jane J Sr, Jane J Jr, Kanev P, Mayes L, Duncan C, Bridgett DJ, Persing JA. The effects of whole-vault cranioplasty versus strip craniectomy on long-term neuropsychological outcomes in sagittal craniosynostosis. Plast Reconstr Surg. 2014 Sep;134(3):491-501. doi: 10.1097/PRS.0000000000000420. PMID: 24804639.
40. Markiewicz MR, Recker MJ, Reynolds RM. Management of Sagittal and Lambdoid

Craniosynostosis: Open Cranial Vault Expansion and Remodeling. Oral Maxillofac Surg Clin North Am. 2022 Aug;34(3):395-419. doi: 10.1016/j.coms.2022.01.005. Epub 2022 Jun 23. PMID: 35752548.

41. Massenburg BB, Tolley PD, Lee A, Susarla SM. Fronto-Orbital Advancement for Metopic and Unilateral Coronal Craniosynostoses. Oral Maxillofac Surg Clin North Am. 2022 Aug;34(3):367-380. doi: 10.1016/j.coms.2022.01.001. Epub 2022 Jul 2. PMID: 35787824.