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# The awareness of treatment plans of craniosynostosis: guideline for patient and family version

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## Abstract

**Background** Functional and structural complexities associated with craniosynostosis present challenges for families of affected individuals. Additionally, these patients undergo childhood surgeries that significantly impact the role of the family/caregiver. Consequently, multiple therapeutic teams have provided various guidelines, with the most recent version in 2023 introducing the patient/family guide. The objective of this study was to evaluate the level of awareness among the patient's family/caregiver regarding the treatment plan.

**Methods** The descriptive-analytical study sample consisted of 117 caregivers (parents) of children diagnosed with craniosynostosis between 2013 and 2023, who were conveniently selected for participation. The self-designed questionnaire was based on the 2023 guidelines and underwent reliability and validity testing. It comprised 14 chapters derived from the literature, with a total of 55 yes/no questions.

**Results** Based on the results obtained from this study, the total correct response rate is 62.3%. The highest level of respondent awareness was associated with Chapter 12, with a correct response rate of 69%. The lowest level of awareness, at 41.8%, was related to Chapter 10.

**Conclusion** It is better to use appropriate educational aids such as educational podcasts based on the latest guidelines for craniosynostosis to educate parents.

**Keywords** Craniosynostosis, Awareness, Treatment plans, Guideline, Patient, Family care

## Background

Care for children with craniosynostosis received attention in the late 1960s [1], and surgical tools and techniques were employed to treat these patients [2]. The best approach for craniosynostosis treatment was introduced

as the collaboration of a multidisciplinary team, with parents of affected children recognized as crucial members of the treatment team [3]. This was because corrective surgery is typically performed in infants, making parents responsible for proxy decisions and active participation in patient care [4].

The first version of guidelines for the care and treatment of craniosynostosis patients was developed by a Dutch team in 2010 and published in 2015 [5]. The second version, endorsed by all participating European societies, was released in 2020 [6]. Subsequently, the latest guidelines for patients and their families, involving European countries and led by the Netherlands, were proposed in 2023. This version consists of 18 chapters providing a simple language explanation of the treatment

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and physical-mental management of craniosynostosis patients for families. However, the healthcare team prioritizes the professional guidelines from 2020 over this version [7].

Although some studies have suggested that caregiver/parental stress can impact the psychological and social outcomes of children with craniosynostosis, no clear trend of increased or decreased stress levels was identified as the cause [8]. On the other hand, most studies indicated that parents were satisfied with the shape of their child's head and surgical outcomes. Some studies also recommended explaining the surgical limitations to parents, and structured interviews can help prepare parents for the desired treatment management [9–11].

In 2020, reports from various countries indicated a prevalence rate of craniosynostosis in children ranging from 4.4 to 7.2 per 10,000 children [12]. Considering the increasing prevalence of craniosynostosis reported in various studies without clear reasons in recent years, it is important to gain a true understanding of the treatment program's awareness, not only among patients but also among their families. Therefore, the present study aims to examine parents' awareness levels regarding their child's craniosynostosis treatment program, using the latest guidelines from 2023.

## Methods

This descriptive-analytical study was conducted in 2023 at Imam Hossein Children's Hospital in Isfahan, Iran. The study sample consisted of 117 caregivers (parents) of children diagnosed with craniosynostosis between 2013 and 2022, who were conveniently selected for participation.

The inclusion criteria for the study were obtaining consent from the caregivers and their ability to be present at the hospital where the surgical procedure was performed. Cases where the caregiver was unable to respond or if the child had passed away for any reason were excluded from the study.

After obtaining ethical codes and necessary permissions, researchers identified the patients and contacted them by telephone.

During these calls, the study objectives were explained, and they were requested to bring their child for examination at the hospital if possible. It was assured that there would be no cost involved for them in this examination. Upon their visit, the caregivers were provided with an informed consent form to sign. In addition to the psychological examination of the patients, the caregivers were interviewed by a trained individual using a self-designed questionnaire. Finally, a total of 91 mothers, 12 fathers, and 14 caregivers (including nurses, grandmothers, and aunts of the child) participated as respondents in the study.

The self-designed questionnaire was based on the 2023 guidelines and underwent reliability and validity testing (see Additional file 1). It comprised 14 chapters derived from the literature, with a total of 56 yes/no questions.

Chapter 1, General Introduction; Chapter 2, Methodology for Guideline Development; Chapter 3, Referral and Diagnostics; Chapter (5 questions) 4, Perioperative Care; Chapter (5 questions) 5, Surgical Treatment of Isolated, Nonsyndromic Craniosynostosis, (7 questions); Chapter 6, Surgical Treatment of Syndromic Craniosynostosis—The Cranial Vault (5 questions); Chapter 7, Surgical Treatment of Syndromic Craniosynostosis—Facial; Chapter (2 questions) 8, Increased Intracranial Pressure (3 questions); Chapter 9, Hydrocephalus (3 questions); Chapter 10, Chiari (1 questions); Chapter 11, Visual, Refractive, And Motility Disorders, (7 questions); Chapter 12, Respiratory Disorders (1 questions); Chapter 13, Hearing Impairments and Speech/Language Development (3 questions); Chapter 14, Dentofacial Abnormalities (5 questions); Chapter 15, (Neuro) Cognitive, Socio-Emotional, And Behavioral Functioning (3 questions); Chapter 16, Psychosocial Functioning (6 questions); Chapter 17, Criteria for Craniosynostosis Expertise Centre and Team Members; Chapter 18, Flow Chart/Patient Summary Visual.

Data were described by descriptive statistics—categorical variables were reported as percentages (%) and frequencies ( $n$ ), while means and standard deviation (SD) were calculated for numerical ones. The level of statistical significance in all used analyses was set at  $p < 0.05$ . Data were processed using the statistical package IBM SPSS V.16.0 (SPSS Inc. Chicago, IL, USA).

## Results

### Socio-demographic and clinical characteristics of the respondents

Ninety-one individuals (8.77%) who responded were mothers. Eighty-two individuals (70%) were boys, and among the children, 34 (1.29%) had metopic craniosynostosis (Table 1).

Table 2 presents the percentage of correct responses for each question. The highest level of respondent awareness was associated with Chapter 12, with a correct response rate of 69%.

The lowest level of awareness, at 41.8%, was related to Chapter 10. The total correct response rate is 62.3%.

The impact of Chapter 8 on overall responsiveness was not statistically significant ( $p = 0.940$ ). Other chapters had a significant impact on overall responsiveness ( $p < 0.05$ ), as demonstrated by the correlation between each chapter and overall responsiveness in Table 2.

Factors influencing the level of awareness of the craniosynostosis treatment program are displayed in Table 3.

**Table 1** Socio-demographic and clinical characteristics of study participants (n = 117)

| Variable                              |                           |                           | Value                  |
|---------------------------------------|---------------------------|---------------------------|------------------------|
| <b>Participants</b>                   |                           |                           |                        |
| Mother, n (%)                         | Age, year, mean ± SD      |                           | 91 (77.8), 28.78 ± 5.5 |
| Father, n (%)                         |                           |                           | 12 (10.2), 32.9 ± 8.47 |
| Caregivers, n (%)                     |                           |                           | 14 (12), 40.14 ± 12.76 |
| Child                                 | Male                      | Age At operation (months) | 82 (70), 7.76 ± 6.77   |
|                                       | Female                    |                           | 35 (30), 8.31 ± 9.1    |
| Operation weight in Kg (mean ± SD)    |                           |                           | 7.24 ± 2.62            |
| Education, n (%)                      | None                      |                           | 45 (38.5)              |
|                                       | Able to read and write    |                           | 62 (53)                |
|                                       | Secondary and preparatory |                           | 7 (6)                  |
|                                       | Tertiary or higher        |                           | 3 (2.6)                |
| Types of craniosynostosis, n (%)      | Sagittal                  |                           | 25 (21.4)              |
|                                       | Metopic                   |                           | 34 (29.1)              |
|                                       | Coronal                   |                           | 12 (10.3)              |
|                                       | Lambdoid                  |                           | 1 (0.9)                |
|                                       | Pansynostosis             |                           | 33 (28.2)              |
|                                       | Multisuture               |                           | 12 (10.3)              |
| Non-syndromic craniosynostosis, n (%) |                           |                           | 47 (40.2)              |
| Syndromic craniosynostosis, n (%)     |                           |                           | 70 (59.8)              |

**Table 2** Awareness of the treatment plan

| Variable, n (%) | 1         | 2         | 3         | 4         | 5         | 6         | 7         | The percentage of correct responses | P      | r     |
|-----------------|-----------|-----------|-----------|-----------|-----------|-----------|-----------|-------------------------------------|--------|-------|
| Chapter 3       | 68 (58.1) | 81 (69.2) | 88 (75.2) | 75 (64.1) | 84 (71.8) |           |           | 58.1                                | 0.006  | 0.255 |
| Chapter 4       | 85 (72.6) | 78 (66.7) | 86 (73.5) | 87 (74.4) | 68 (58.1) |           |           | 59.8                                | 0.002  | 0.288 |
| Chapter 5       | 68 (58.1) | 60 (51.3) | 78 (66.7) | 83 (70.9) | 86 (73.5) | 80 (68.4) | 75 (72.6) | 62.4                                | 0.001  | 0.433 |
| Chapter 6       | 85 (72.6) | 84 (71.8) | 74 (63.2) | 79 (67.5) | 81 (69.2) |           |           | 62.9                                | 0.0001 | 0.330 |
| Chapter 7       | 75 (64.1) | 71 (60.7) |           |           |           |           |           | 60.1                                | 0.013  | 0.229 |
| Chapter 8       | 75 (64.1) | 75 (64.1) | 71 (60.7) |           |           |           |           | 56.1                                | 0.940  | 0.007 |
| Chapter 9       | 66 (56.4) | 76 (65)   | 68 (58.1) |           |           |           |           | 57.7                                | 0.031  | 0.199 |
| Chapter 11      | 64 (54.7) | 64 (54.7) | 61 (52.1) | 65 (55.6) | 74 (63.2) | 80 (68.4) | 65 (55.6) | 67.7                                | 0.0001 | 0.430 |
| Chapter 10      | 51 (43.6) |           |           |           |           |           |           | 41.8                                | 0.001  | 0.987 |
| Chapter 12      | 68 (58.1) |           |           |           |           |           |           | 69                                  | 0.0001 | 0.335 |
| Chapter 13      | 72 (61.5) | 66 (56.4) | 73 (62.4) |           |           |           |           | 68.9                                | 0.002  | 0.286 |
| Chapter 14      | 69 (59)   | 73 (62.4) | 72 (61.5) | 68 (58.1) | 67 (57.3) |           |           | 59.6                                | 0.0001 | 0.389 |
| Chapter 15      | 69 (59)   | 67 (57.3) | 61 (52.1) |           |           |           |           | 64.7                                | 0.0001 | 0.351 |
| Chapter 16      | 78 (66.7) | 72 (61.5) | 70 (59.8) | 71 (60.7) | 67 (57.3) | 78 (66.7) |           | 63                                  | 0.002  | 0.286 |

PP-value, r correlation

An important finding is that, within the education model, the type of craniosynostosis had a significant impact on the respondent’s awareness of the treatment program ( $p=0.009$ ), while the respondents’ education level was not significant.

In the age model of the respondents, neither the type of craniosynostosis child ( $p=0.464$ ) nor the respondents’ age had a significant impact on the level of awareness of

the treatment program. Similarly, within the respondent’s familial relationship model with the child, neither the type of craniosynostosis child ( $p=0.500$ ) nor the respondents’ familial relationship had a significant impact on the level of awareness of the treatment program.

In the syndromic craniosynostosis model, neither the type of craniosynostosis child ( $p=0.785$ ) nor the

**Table 3** Factors associated with awareness of the treatment plan from multiple logistic regression models

| Variable                            | Awareness of the treatment plan |       |
|-------------------------------------|---------------------------------|-------|
|                                     | OR <sup>a</sup> (95% CL)        | P     |
| Age (participants) (Ref < 30 years) | 1.006 (0.914–1.108)             | 0.592 |
| Education Status (Ref: none)        |                                 |       |
| Able to read and write              | 1.069 (0.960–1.191)             | 0.225 |
| Secondary and preparatory           | 1.037 (0.838–1.284)             | 0.736 |
| Tertiary or higher                  | 0.823 (0.601–1.127)             | 0.224 |
| Participants (Ref: mother)          |                                 |       |
| Father                              | 1.003 (0.856–1.174)             | 0.973 |
| Caregivers                          | 0.927 (0.799–1.076)             | 0.318 |
| Craniosynostosis (Ref: syndromic)   |                                 |       |
| Non-syndromic craniosynostosis      | 0.986 (0.895–1.086)             | 0.770 |

<sup>a</sup> ORs are adjusted for all the variables in the first column of this table

**Table 4** Factors associated with awareness of treatment plan (chapters) from multiple logistic regression models

| Variable                     | Awareness of the treatment plan |       |
|------------------------------|---------------------------------|-------|
|                              | OR* (95% CL)                    | P     |
| Chapter 11                   |                                 |       |
| Participants (Ref: mother)   |                                 |       |
| Father                       | 0.099 (0.574–1.738)             | 0.997 |
| Caregivers                   | 0.575 (0.343–0.964)             | 0.036 |
| Chapter 12                   |                                 |       |
| Participants (Ref: mother)   |                                 |       |
| Father                       | 1.025 (0.273–3.851)             | 0.971 |
| Caregivers                   | 12.941 (1.452–115.302)          | 0.022 |
| Chapter 8                    |                                 |       |
| Education Status (Ref: None) |                                 |       |
| Able to read and write       | 1.373 (0.795–2.372)             | 0.256 |
| Secondary & Preparatory      | 1.110 (0.121–10.203)            | 0.926 |
| Tertiary or Higher           | 4.744 (1.132–19.888)            | 0.033 |

\*OR Odds Ratio

syndromic/non-syndromic craniosynostosis had a significant impact on the level of awareness of the treatment program.

In Table 4, all variables were examined individually with each chapter. The results indicated that in the 11th and 12th chapters, the caregiver’s level of awareness had a significant difference compared to the father with a higher odds ratio. Additionally, in Chapter 8, the level of awareness among educated individuals with a higher odds ratio displayed a significant difference compared to individuals with lower levels of education.

In this study, 73% of caregivers stated that they have used social media platforms such as Instagram, along with the physician’s explanations, to increase their level of awareness. Additionally, 52% of caregivers reported using information from other parents of affected children, alongside the physician’s explanations.

### Discussion

Based on the results obtained from this study, the highest level of respondent awareness was associated with Chapter 12, with a correct response rate of 69%. The lowest level of awareness, at 41.8%, was related to Chapter 10. Total correct response rate of 62.3%.

A comprehensive study on the awareness level of caregivers of children with craniosynostosis regarding their treatment program was not found. However, Kajdic (2018) [13] emphasized the vital importance of early diagnosis, surgical techniques, postoperative care, and sufficient follow-up in the treatment of craniosynostosis in a qualitative study. Furthermore, untreated craniosynostosis can lead to serious complications such as growth retardation, facial abnormalities, sensory, respiratory, and neurological disorders, as well as visual impairments and psychological disturbances. As observed, the caregivers’ awareness level of these disorders is above 60%, indicating the need to incorporate effective educational strategies into the patient’s treatment program to enhance this awareness.

Anantheswar (2009) [14] stated that parental counseling is an important and integral part of the treatment of children with craniosynostosis. Recent advancements in surgical techniques and the concept of a family-centered approach have significantly improved the safety and treatment outcomes for these children. In the present study, the caregivers’ awareness of the treatment plan was found to be 62.3%.

Lun (2022) [15] mentioned that one of the concerns of parents is the shape of their child’s head. In the current study, 73.5% of the caregivers expressed that the abnormal shape of the skull improves spontaneously after surgical intervention.

Jong (2011) [16] stated that most patients with craniosynostosis have syndromic and complex forms, accompanied by recurrent otitis media with effusion, which leads to a reduction in conductive hearing throughout their lives. The awareness level of the caregivers regarding this hearing impairment was obtained as 68.9% in the present study.

The results of the study by Liasis (2011) [17] indicated that all children with craniosynostosis had visual field defects compared to other children. Hinds (2022) [18] also showed a high prevalence of amblyogenic factors in patients with craniosynostosis. Duan (2021) [19]

mentioned that a pediatric ophthalmologist is an essential part of the multidisciplinary care team, and their continuous follow-up can help minimize the risk of vision impairment in craniosynostosis patients. The results of a systematic review also indicated a high prevalence of ocular abnormalities in both syndromic and nonsyndromic craniosynostosis. All studies were aligned with Chapter 11, and the respondents' level of awareness was 67.7%.

Choi (2016) [20] stated that intracranial pressure, hydrocephalus, Chiari malformation, and neuropsychological dysfunction are major concerns in pediatric neurosurgery for children with craniosynostosis, all of which are addressed in the 2023 guidelines. In the present study, it was found that caregivers had a higher awareness of neuropsychological dysfunction and a lower awareness of Chiari malformation.

One limitation of this study is that it would have been better to first ask the caregivers questions and then provide them with the necessary training based on the 2023 guidelines. Subsequently, the same questions could have been asked again to measure the impact of these guidelines on caregiver awareness. Therefore, it is suggested that further studies be conducted in this regard.

## Conclusions

Overall, it can be concluded that caregivers of children with craniosynostosis had above-average awareness of all the questions related to each chapter. In this regard, further explanations in simple language should be provided to the parents or caregivers by the treating physician and nurse. Additionally, appropriate educational aids such as educational podcasts based on the latest guidelines should be utilized. Furthermore, considering that caregiving approaches may vary depending on the type of craniosynostosis and accompanying syndromes, parents/caregivers should be well-informed about the type, cause, and treatment of the patient.

## Supplementary Information

The online version contains supplementary material available at <https://doi.org/10.1186/s43054-024-00273-3>.

**Additional file 1.** Questionnaire.

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## Ethical issues

Written consent was obtained from the families of patients to enter this study.

## Authors' contributions

All authors read and approved the final manuscript. BA contributed as the main author with the concept of planning the study. AM, AR, and DSHT

contributed to study design, patient selection, and follow-ups. SF and DSHT performed the statistical analysis, interpreted the data, and helped write the manuscript. BA and AR mentored the edition of the final version.

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## Availability of data and materials

The data that support the findings of this study are available but restrictions apply to the availability of these data, which were used under license for the current study, and so are not publicly available. Data are however available from the authors upon reasonable request and with permission of [DSHT].

## Declarations

### Ethics approval and consent to participate

The current study was approved by the Isfahan University of Medical Sciences Ethics Committee with the code of IR.MUI.RESEARCH.REC.1399.051.

### Consent for publication

All coauthors have seen and agree with the contents of the manuscript and each author believes that the manuscript represents honest work. All coauthors certify that the submission is not under review at any other publication. There are no previous reports that might be regarded as redundant publications of the same or very similar work. Furthermore, the authors report no conflict of interest.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

The study was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

### Competing interests

The authors declare that they have no competing interests.

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