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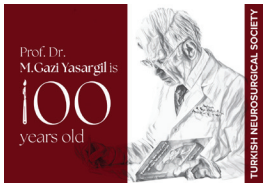

Non-Surgical Management of Trigonocephalic Patients: An OCT and 3D-CT Based Follow-up Study

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ABSTRACT

AIM: To evaluate the cosmetic and visual aspects of trigonocephaly patients who have not received surgical intervention throughout a long-term follow up.

MATERIAL and METHODS: Patients with trigonocephaly who did not undergo surgery were evaluated. Using 3D computed tomography images, the frontal (angle of the 2 lines between the bilateral pterion and nasion) and interfrontal angles were measured, and the frontal projections at the first and last examinations were compared. The final appearance was photographed, and detailed eye examinations and optical coherence tomography (OCT) measurements were performed.

RESULTS: The study included 6 patients (3 male and 3 female patients). The patient age at diagnosis was between 3 months and 2 years. The mean age at diagnosis was 10.33 months, and the average follow up period was 34 months. The interfrontal angle was not below 118° in any of our patients. We noted that the interfrontal angle increased with an increase in the follow up period. The ophthalmologic examination differed according to the age of the patients. Extraocular motility testing revealed no abnormalities. The anterior segment and fundus examination results were within normal limits. Bilateral OCT images of the optic discs revealed normal retinal nerve fiber layer (RNFL) thickness. No problems were detected in the neurocognitive development of the patients, and no severe cosmetic pathology was observed.

CONCLUSION: Neurocognitive retardation and ophthalmologic problems were not detected in our patients with trigonocephaly who did not undergo surgery. More detailed studies with larger sample sizes are required before changing our approach to the treatment of trigonocephaly.

KEYWORDS: Craniosynostosis, Trigonocephaly, Unoperated

ABBREVIATIONS: CT: Computed tomography, **OCT:** Optical coherence tomography, **RNFL:** Retinal nerve fiber layer

INTRODUCTION

Trigonocephaly is a type of craniosynostosis that involves the premature closure of the metopic suture. Single suture synostoseis is encountered of every 1 in 2000 live births, and trigonocephaly accounts for 10%-15% of all craniosynostoseis cases (11). Trigonocephaly can be mild, moderate or severe, and the mild forms may not be distinguishable from the metopic ridge (3).

Craniosynostosis is not only a cosmetic anomaly but also an important problem that restricts brain development. For anterior synostosies, the rate of mental deficit was shown to be 31.6% among those who did not undergo surgery during

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the first year of life, when the brain develops most rapidly (7). In cases of simple craniosynostosis, an increase in intracranial pressure of approximately 13% can be observed. Stasis papillae are seen at the fundus of the eye in 16%-25% of cases involving increased intracranial pressure (1).

Early surgery is recommended because of the possibility of serious neurocognitive problems if treatment is delayed; however, some families reject surgery. This study aimed to evaluate the cosmetic and visual aspects of trigonocephaly in patients who did not undergo surgery during at long-term follow up because their families rejected this treatment option.

MATERIAL and METHODS

The study included, patients with trigonocephaly who did not undergo surgery. Using three dimensional (3D) computed tomography (CT) images, the frontal (angle of the 2 lines between the bilateral pterion and nasion) and interfrontal angle were measured, and the frontal projections at the first and last examinations were compared. The presence of a Chiari malformation was assessed. The final appearance was photographed, detailed eye examinations were performed, and optical coherence tomography (OCT) measurements were performed (4 of 6 patients). The study protocol was approved by the local ethics committee (approval number: 2024-TBEK 2024/02-08), and informed consent forms were signed by the guardians of the patients.

RESULTS

The study included 6 patients with trigonocephaly 3 male and 3 female. The patient age at diagnosis was between 3 months and 2 years. The mean patient age at diagnosis was 10.33 ± 7.39 months (range: 3 - 24 months). The follow up period of the patients ranged from 9 to 69 months, with an average of 34 months. The frontal angle was measured retrospectively from the cranial CT images of the patients obtained at the time of diagnosis. An angle of 89° was considered to indicate severe trigonocephaly, 90°-95° was considered to indicate moderate trigonocephaly, and 96°-103° was considered to indicate mild trigonocephaly. Accordingly, based on frontal angle measurements, 1 patient had mild, 1 had moderate trigonocephaly, and 4 had severe trigonocephaly. The interfrontal angle was not below 118° in any of the patients. However, the interfrontal angle increased from the time of diagnosis to the last follow up. The angle increased by 15.1° at 69 months of follow up, 10.8° at 42 months, 8.8° at 34 months, 8.8° at 27 months, 16.7° at 23 months, and 4.1° at 9 months. The greatest increase in the angle was found in our patient who had the lowest interfrontal angle and was followed between the ages of 2 years and 3 years and 11 months. However, in general, the interfrontal angle increased with an increase in the follow up period (Figure 1, Table I).

In patients 1, 2, 3, and 5, ocular examinations revealed bilateral best-corrected visual acuity of 20/20, with normal light responses. In these patients, extraocular motility testing revealed no abnormalities, and the anterior segment and fundus examination results were within normal limits. Moreover, bilateral OCT images of the optic discs revealed normal retinal nerve fiber layer (RNFL) thickness (Figure 2). The average RNFL thickness in these 4 patients was 101.875 ± 4.356 µm. In patients 4 and 6, the anterior segment and fundus examination results were within normal limits (Table II). In these patients, normal light responses were observed bilaterally. Due to the young age of these patients, visual acuity could not be assessed, and OCT imaging could not be performed. However, the fixation and object-tracking abilities of the patients were good. Moreover, there were no abnormalities in the extraocular motility test.

Chiari malformation was not detected in any of our patients (Table I). Moreover, no problems were detected in the neurocognitive development of the patients, and no severe cosmetic pathology was observed on the macroscopic examination of the patients (Figure 3).

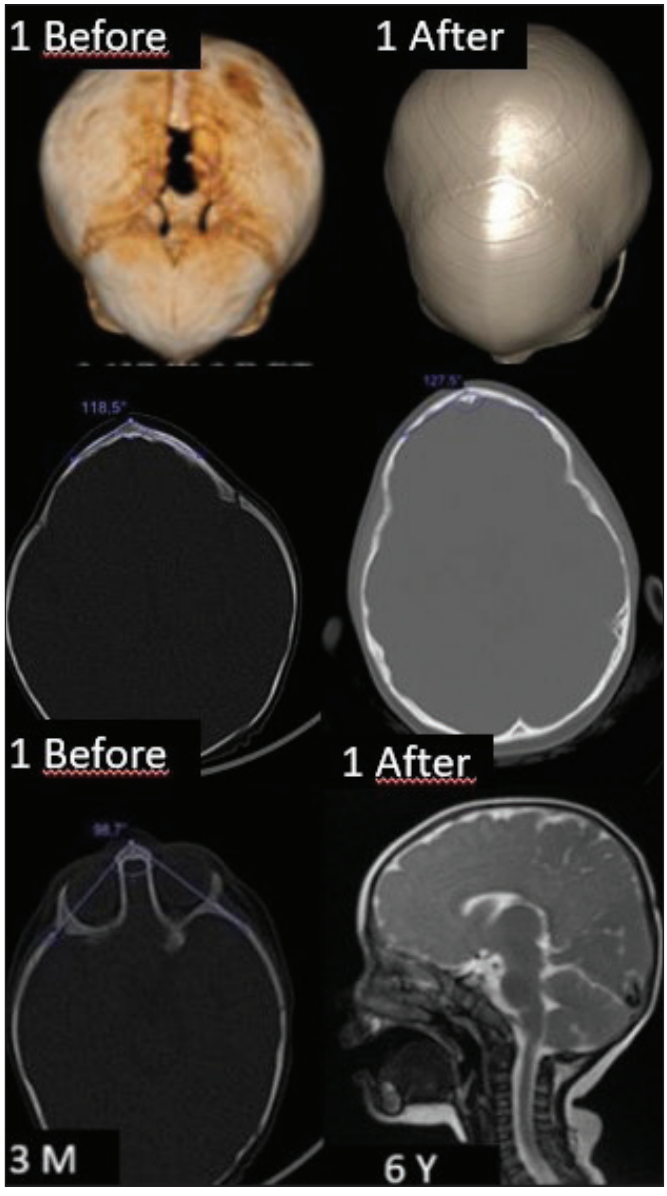

Figure 1: Measurement and radiological examinations of a patient 3 months and 6 years of follow-up (M: months, Y: year).

Table I: Patients' frontal and Interfrontal Angles and the Presence of Chiari Malformation

Patients-Follow / Measurement	Frontal angle	Interfrontal angle (Diagnosis time)	Interfrontal angle (Check time)	Chiari Malformation
1 (3-72 months)	98.7	131.6	146.7	-
2 (10-48 months)	92.5	137.8	146.6	-
3 (12-46 months)	78.4	137	145.8	-
4 (6-33 months)	87.5	139.5	146.3	-
5 (24-47 months)	86	123.5	140.2	-
6 (7-16 months)	82.2	127.7	131.8	-

Table II: OCT Measurements of the Patients

RNFL thickness/Case No	1/R	1/L	2/R	2/L	3/R	3/L	5/R	5/L	Average	Standard Deviation
Average	109	107	99	96	102	99	103	100	101.875	4.356
Superior	138	131	116	119	124	116	104	126	121.75	10.43
Inferior	127	134	126	111	123	128	158	123	128.75	13.498
Temporal	70	77	70	74	83	83	79	77	76.625	5.097
Nasal	102	85	85	81	78	69	71	73	80.5	10.609
Superior Temporal	128	122	115	115	138	124	121	140	125.375	9.47
Superior Nasal	147	141	116	124	109	108	88	113	118.25	18.956
Nasal superior	105	91	92	89	88	70	75	72	85.25	11.973
Nasal inferior	99	79	78	74	69	68	67	74	76	10.309
Inferior nasal	135	116	108	94	91	120	133	86	110.375	18.844
Inferior temporal	118	151	144	129	155	135	184	160	147	20.479
Temporal inferior	60	85	64	76	90	66	78	74	70.125	10.398
Temporal superior	80	69	76	71	76	100	80	79	78.875	9.448

DISCUSSION

The metopic suture starts to fuse in the 3rd month and closes completely in the 8th month (9). It constitutes 10%–15% of all craniosynostosis cases and 25% of all nonsyndromic craniosynostosis cases (14). Di Rocco et al. reported that the incidence of trigonocephaly has increased by 420% in the last 20 years (4).

Brain development, movement ability, and language development occur in the first 2 years of life. Although almost all children with single-suture synostosis can lead normal lives, among all craniosynostosis cases, trigonocephalic cases involve the most neurocognitive developmental delay (9). It has been reported that corpus callosum anomalies are common in patients with trigonocephaly. Dyslexia and language-speech disorders have been observed due to cerebellar pathologies resulting from single-suture synostosis (2).

A decrease in frontal lobe blood flow is observed in patients with preoperative trigonocephaly under 18 months of age. Surgery is indicated in cases of trigonocephaly, especially if findings of increased intracranial pressure are detected and there is a decrease in frontal lobe perfusion (8). Corrective surgery for trigonocephaly performed before 12 months of age was reported to prevent an increase in intracranial pressure, but despite surgery, neurological and behavioral disorders and visual problems may still occur in patients (13).

Many studies in the literature have performed, intensive evaluations of neurocognitive development in patients with trigonocephaly. In our study, no neurocognitive developmental delay was detected in examinations or family observations. Due to the retrospective nature of the study, it is impossible to say this is the best neurocognitive outcome. We would like to point out that patients who do not undergo surgery can be educated without any problems.

Skull deformity, which is generally noticed at birth, becomes more evident between the ages of 6 and 9 months. During infancy, head deformity is more obvious because the head constitutes 25% of the overall body structure. In adults, the

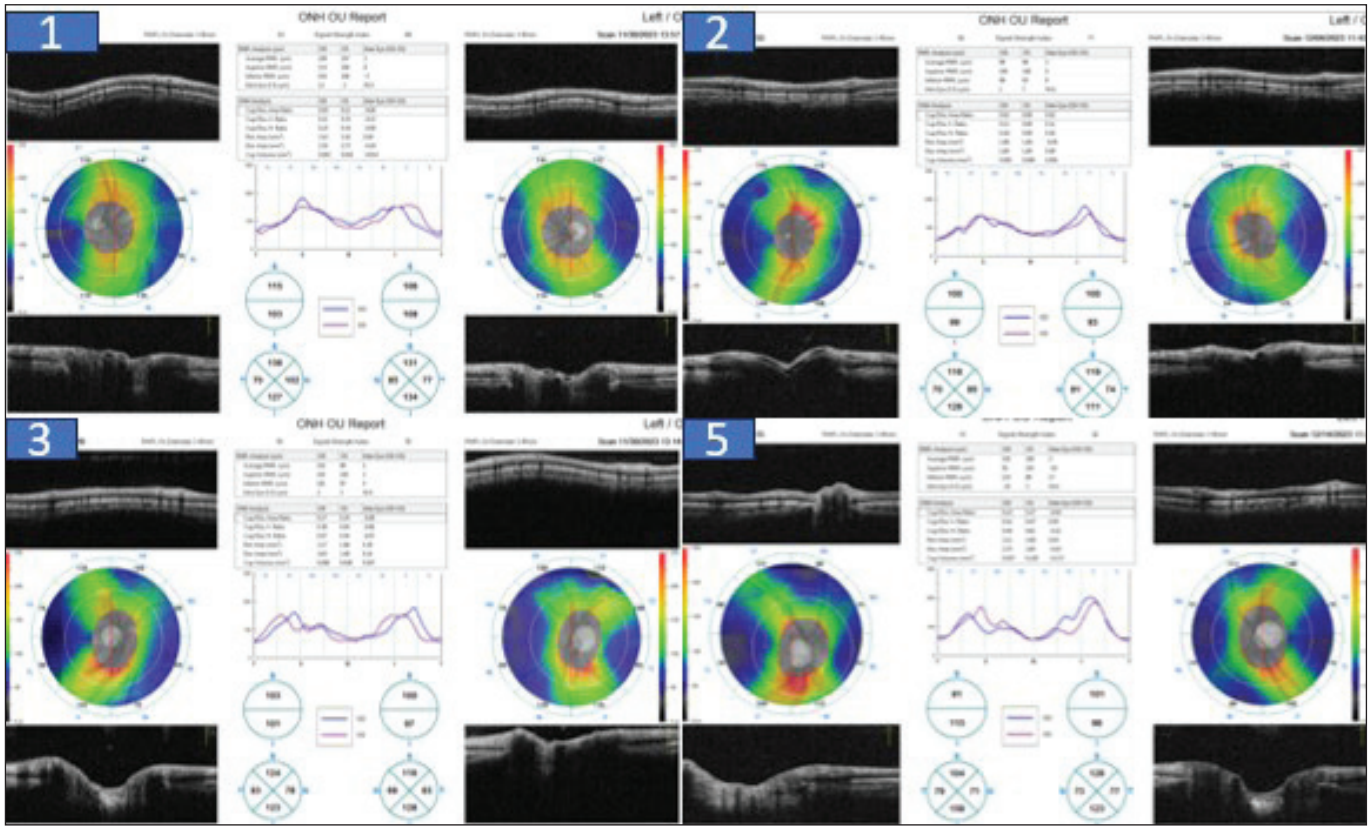

Figure 2: OCT examinations of 1st, 2nd, 3rd and 5th cases.

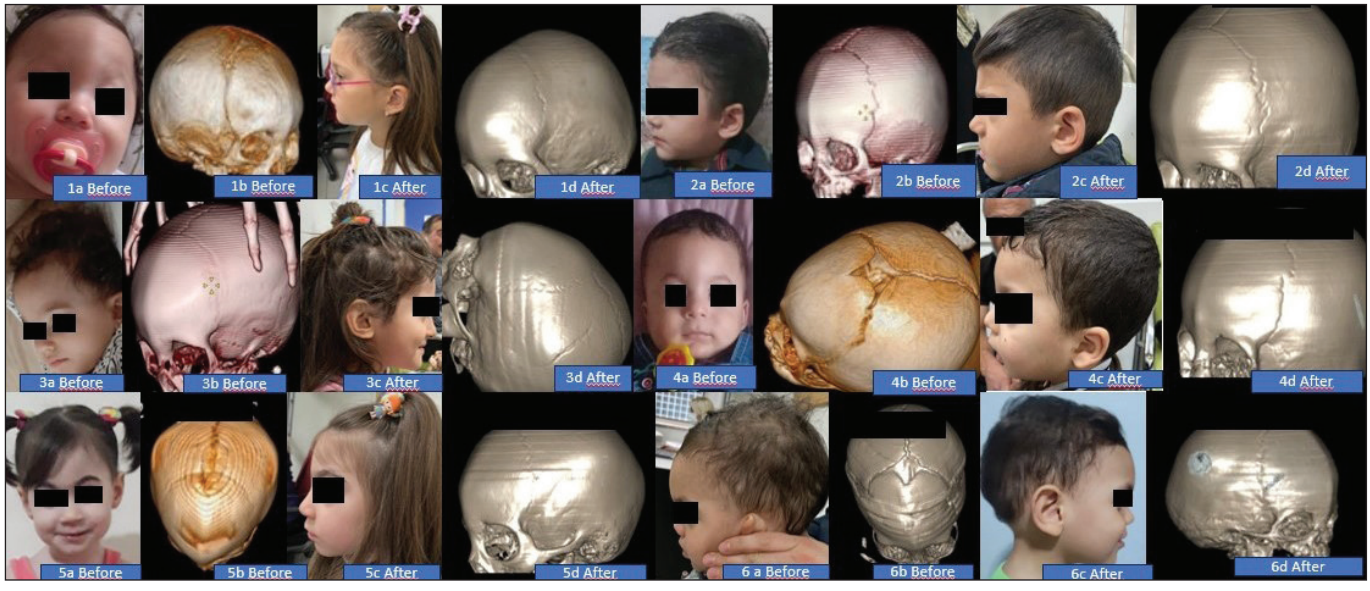

Figure 3: Photographic and radiological follow-up of patients (Before and after).

head-to-body ratio is 9%–10%, and thus, deformities are less noticeable (9). In our study, head shape anomalies improved significantly as the follow up period and patient age increased.

Although many ophthalmological problems occur in patients with craniosynostosis, papilledema and amblyopia must be controlled and treated to avoid a reduction in visual acuity (12). In our study, eye examinations were performed in all patients, and no obvious pathology was detected. OCT is known to be important for detecting elevated intracranial pressure in patients with craniosynostosis (10). In our study, OCT was performed in 4 of the 6 patients, and the results were within normal limits. We identified 3 studies about RNFL thickness in healthy children. In the first study, the average RNFL thickness measured by OCT in healthy preschool children (age 3–6 years) was 105.5 ± 11.5 μm (15). In the second study, the average RNFL thickness in 6-year-old children was 103.7 ± 11.04 μm (6). In the third study, the average RNFL thickness in children aged 3–17 years was 96.49 ± 10.10 μm (5). In our study, the average RNFL thickness was 101.875 ± 4.356 μm. No significant differences were found when our results were compared with those of the 3 studies.

CONCLUSION

In our study, contrary to current knowledge and studies in the literature, cosmetic appearance (head shape anomalies) and ophthalmological problems were not detected in patients with trigonocephaly who did not undergo surgery. More detailed studies with larger sample sizes are required before changing our approach to the treatment of trigonocephaly.

Declarations

Funding: There was no commercial or private funding.

Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

Disclosure: The authors declare no competing interests.

AUTHORSHIP CONTRIBUTION

Study conception and design: MOT, EBG

Data collection: EBG

Analysis and interpretation of results: EBG, FY

Draft manuscript preparation: EBG, MOT

Critical revision of the article: EBG, MOT

Other (study supervision, fundings, materials, etc...): EBG, MOT

All authors (EBG, FY, MOT) reviewed the results and approved the final version of the manuscript.

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