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ORIGINAL PAPER

The metopic-sagittal craniosynostosis—report of 35 operative cases

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Abstract
Purpose We have diagnosed 35 cases of the supposedly rare condition metopic-sagittal synostosis in the past 20 years. Here, we introduce their clinical symptoms, neuroradiological findings, and surgical treatment methods, as well as discuss the relevant literature.
Methods Subjects included 35 patients (33 boys and 2 girls; mean age 4.2 years; range 1–8 years). Magnetic resonance imaging (MRI) confirmed that there were no abnormal findings in the brain. Thirty patients presented with symptoms including speech delay, hyperactivity, autistic tendency, motor impairment, self-mutilation, and panic/temper tantrum behaviors. No other congenital malformation was observed, and all cases were considered to be the non-syndromic type. The final diagnosis was made using three-dimensional computed tomography (3D-CT) scans. The surgery was done the fronto-orbital advancement in addition to remove the large parts of sphenoid bones including sphenoid ridges at the skull base and trimmed the calvarium as necessary to reduce pressure.
Results Surgical intervention improved clinical symptoms in nearly all 35 patients; cosmetic problems in patients with scaphocephaly were also corrected.
Conclusions In the cases of child patients with metopic-sagittal synostosis who had clinical symptoms, surgical intervention improved such symptoms, suggesting its potential utility for metopic-sagittal synostosis with clinical symptoms. A surgical procedure focusing on the skull base was important for our successes. Based on the fact that metopic-sagittal synostosis was diagnosed in 35 patients at one institution over a relatively short period of time, this pathological condition may not be as rare as is currently believed.

Keywords Metopic-sagittal synostosis · Developmental delays · Mild trigonocephaly · Sphenoid ridge

Introduction

We have dealt with cases of mild trigonocephaly exhibiting various clinical symptoms since 1994 [20–23]; within these cases, we diagnosed cases where the metopic suture is fused with the sagittal suture, a phenomenon known as metopic-sagittal synostosis. This pathological condition is currently considered to be rare [3].

Currently, we have diagnosed 35 patients with metopic-sagittal synostosis. Patients presented with clinical symptoms that were improved to varying degrees following surgery in almost all cases.

No study of a large number of patients with this pathological condition has been reported, but the frequency of such patients being seen by our hospital suggests there may be many more cases than conventionally believed. Therefore, to improve the diagnosis and treatment of metopic-sagittal synostosis, we report here the analysis of clinical symptoms, neuroradiological diagnostic procedures, and surgical procedures and outcomes in our patients.

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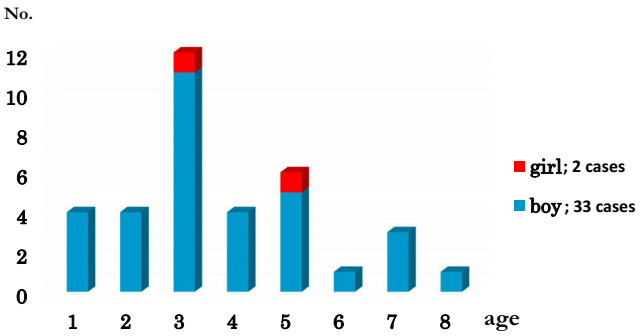


Fig. 1 Age and sex distribution of the study population

Methods

Subjects

The subjects were a total of 35 patients who were diagnosed with metopic-sagittal synostosis: 16 patients at Okinawa Prefectural Naha Hospital between November 1997 and March 2006 and 19 patients at Okinawa Prefectural Nanbu Medical Center/Nanbu Child Medical Center (formerly Okinawa Prefectural Naha Hospital) between April 2006 and January 2016. The subjects included 33 boys and 2 girls, and their ages ranged from 1 to 8 years (mean age = 4.2 years) (Fig. 1).

All the patients underwent chromosome testing, but no abnormal results were found. No other congenital malformation was observed, and all cases were considered to be the non-syndromic type.

In the family histories, two male siblings related to each of three patients and a female cousin of one patient had undergone surgery for mild trigonocephaly.

Clinical symptoms

All patients received a full clinical work-up that included neurological and psychiatric tests to assess behavior, cognition, and motor function and to detect the presence of specific or global developmental delays. In our assessments, we found no developmental delays in five patients. In 30 patients, we noted various symptoms; speech delay was the primary developmental delay. Speech delay and subsequent improvement were assessed according to the degrees of verbal capability established by the National Rehabilitation Center Sign-Significance Test (NRC S-S test) [12].

Diagnosis

A physical examination and visual inspection was used to initially diagnose the patients; subjects were inspected for ridges at the metopic and sagittal sutures either visually or by palpation. The diagnosis was confirmed by using a

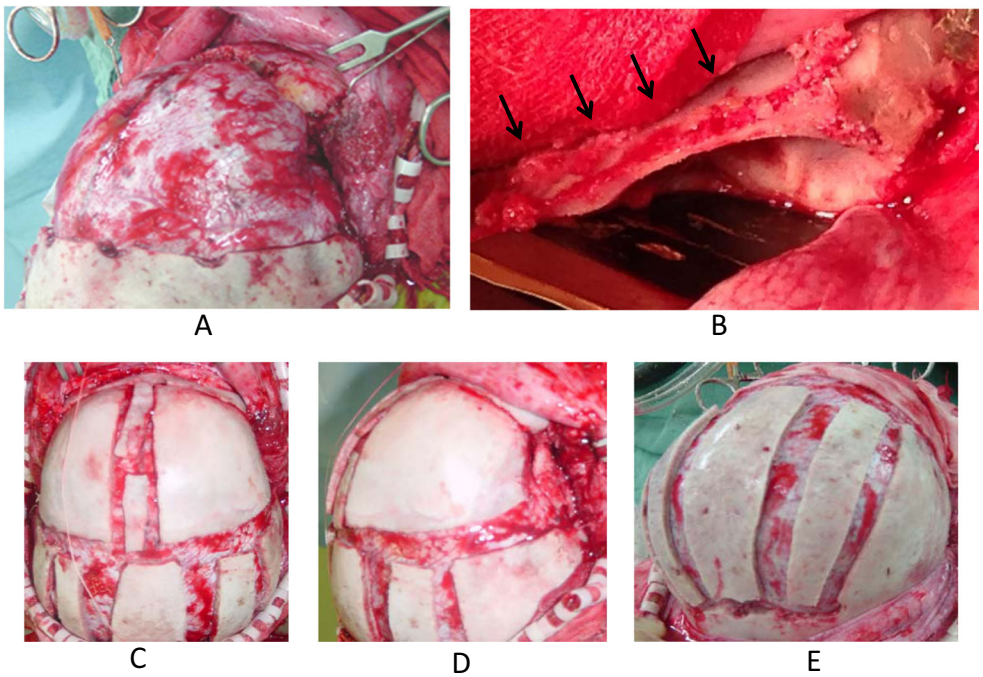


Fig. 2 Surgical views and 3D-CT imaging findings immediately following surgery. **a** After removing the frontal calvarium and orbital bar. **b** The very wide sphenoid ridge (*arrows*) was removed up to the meningo-orbital band. **c**, **d** After the posterior craniectomies, pieces of bone were replaced in floating fashion, and the orbital bar was sutured to the lateral orbit. **e** The posterior craniectomies were done on the bilateral parietal bones. The most posterior points were several centimeters posterior to lambda. **f** 3D-CT imaging immediately following surgery for the scaphocephalic type. **g** 3D-CT imaging immediately following surgery for the non-scaphocephalic type

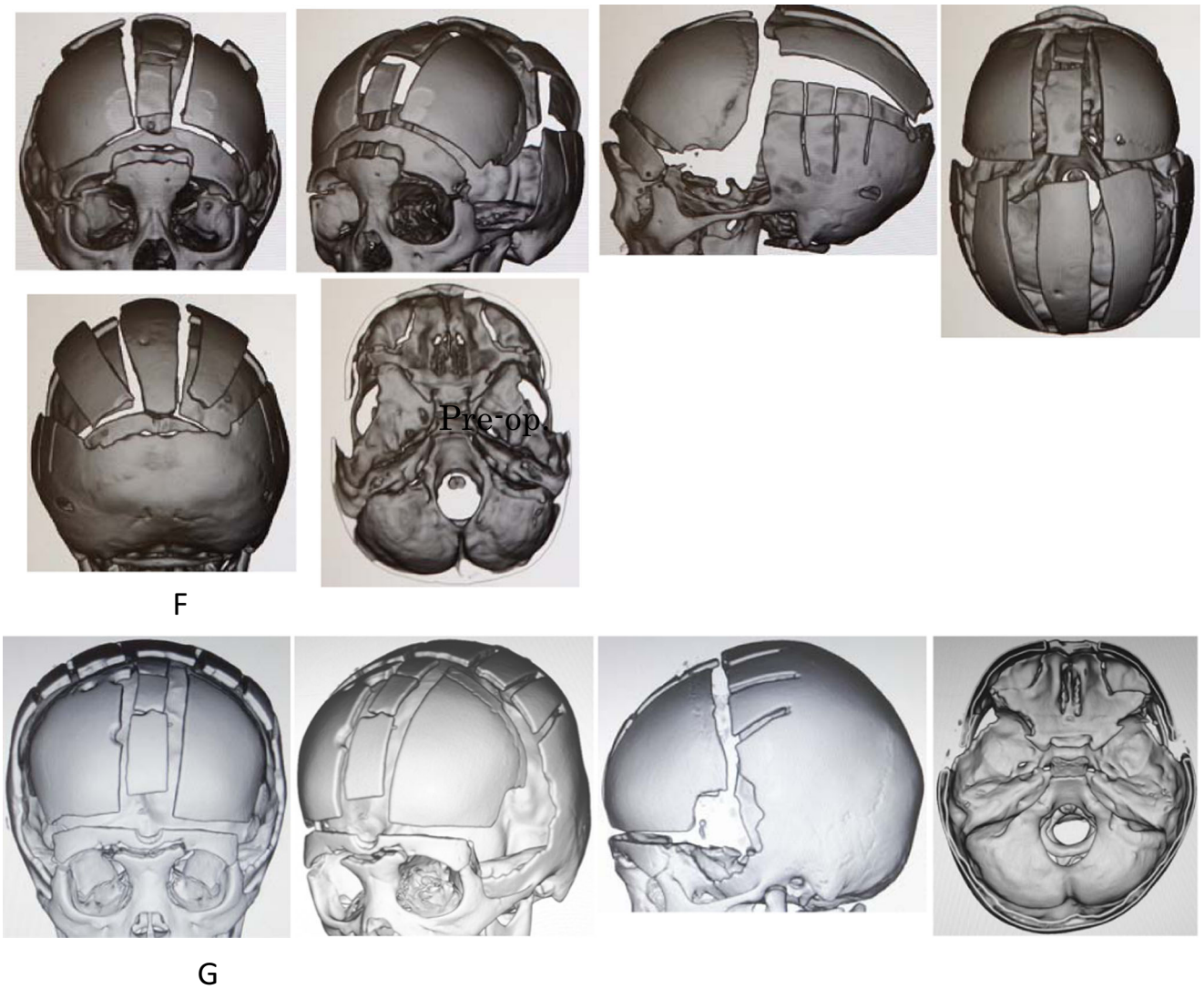


Fig. 2 continued.

three-dimensional computed tomography (3D-CT) scan to confirm fusion of the metopic ridge and sagittal suture, as well as to visually confirm a sagittal ridge first identified by palpation. We used MRI to confirm that no congenital malformations or abnormal structural problems were present. Finally, we obtained plain skull X-rays before surgery in all patients to verify that the metopic and sagittal sutures were closed.

Monitoring of intracranial pressure

Intracranial pressure (ICP) was measured during surgery in 28 patients. After the scalp was flipped, a burr hole was prepared in the forehead and a sensor (Camino's Monitoring System, Integra Lifesciences Corporation, NJ, USA) inserted into the epidural space. PCO_2 was held at approximately 30 mmHg for the first measurement and 38–42 mmHg for the second, during which measurements were taken for several minutes.

Surgical procedure

Patients underwent one or both of the procedures described below, depending on their diagnoses. The anterior procedure was only performed in the patients with the non-scaphocephalic type; additionally, some fracture lines were introduced into the parietal bone (Fig. 2). The patients with the scaphocephalic type underwent both the anterior and posterior procedures.

Anterior We performed a bifrontal craniotomy in which the open-end was approximately 3 cm posterior to the coronal sutures. The frontal calvarium was extracted as one piece. Next, the lesser and greater wings of the

Table 1 Clinical characteristics and post-surgical outcomes in the patient group

	Case	Sex	Age at surgery (years)	Date of operation	Pre-op symptoms and signs	Digital markings	Mean ICP (PCO_2)	Changes in symptoms and signs after surgery	Period of follow-up (years)	Age at present (years)	Status at present
Type 1	1. K.Y.	Boy	1.7	May 17, 1999	No meaningful words Poor understanding Hyperactive Unstable walk Self-mutilation	4	N/A	Acquisition of more than 10 words after 3 months Greatly reduced hyperactivity Stable walk Eliminated self-mutilation Spoke full sentences after 3 years in the SE class of primary school at age 6 with normal conversational ability	16.8	18.5	Graduated SE high school; employed with mildly impaired intelligence
	2. M.T.	Boy	3.1	January 13, 2000	Unsteady walk No meaningful words	4	N/A	Steady walk after 6 months Few words after 1 year Attending SE primary school Age 6 DQ = 43	16.2	19.3	Attending SE high school Engages in simple conversation
	3. N.Y.	Boy	5.2	January 11, 2000	3-word sentences with difficulty, including poor pronunciation Hyperactive	4	8 (38)	Speaking longer sentences after 1 month Greatly reduced hyperactivity At age 6, in regular primary school; some difficulties in schoolwork in the third grade DQ = 62	15.2	20.4	No problems in conversation Graduated an SE junior high school; engaged in a part time job Good interpersonal relationships; mildly impaired intelligence
	4. U.R.	Boy	3.9	March 26, 2001	Several words Could not play with other children	4	6 (39.9)	Improved facial expression Increased vocabulary; playing with other children after 1 year Better understanding Uses two-word sentences and converses with his mother after 2 years	15	18.9	Graduated SE high school in March 2016; employed at a nursing home Holds natural conversations; mildly impaired intelligence
	5. K.K.	Boy	4.1	July 1, 2002	2-word sentences but difficulties in conversation Hyperactive Poor pronunciation DQ = 77 (as of February 12, 2011)	4	12 (36.9)	Improved language acquisition and understanding Greatly reduced hyperactivity after 6 months DQ = 89 after 6 months DQ = 99 after 18 months Entered primary school at age 6	13.7	17.8	Unknown
	6. U.S.	Girl	2.2	August, 12, 2002	Delayed language development 2-word sentences; frequent temper tantrums	4	19 (36.2)	Language capabilities almost normal and no temper tantrums after 4 months DQ = 83 at age 3 In regular primary school at age 6; has many friends	13.6	15.8	High school in a special advanced class
	7. Y.M.	Boy	5.2	February 24, 2003	3-word sentences but difficulties in conversation Panic Head banging; antisocial behavior	4	11 (32.9)	–	13	18.2	Graduated an SE high school; holds simple conversations Working in a disability frame No problems in his daily activities
	8. O.Y.	Boy	6.6	April 14, 2003	Nonstop talking resulting in 1-way street conversations Head banging Hyperactive Impatience	4	10 (42.8)	No changes after 2 years but enrolled in the SE class of primary school Symptoms gradually improved over the years; graduated from a commercial high school.	13	19.6	Difficulties in communication with others but no problems in daily conversation; actively job-hunting
	9. O.T.	Boy	3.6	December 20, 2004	10-word vocabulary Hyperactivity Diagnosed with autism spectrum disorder Difficulty making eye contact and playing with other children Other-inflicted	3	N/A	2-word sentences Increased acquisition of language Other-inflicted ceased Reduced hyperactivity after 6 months Improved eye contact and sociability	11.3	14.9	In third grade of SE junior high school Echolalic speech Severe autism disorder
	10. T.I.	Boy	3.7	December 12, 2005	No meaningful words	2	14 (38.1)	Vocabulary developing after 18 months >4-word sentences after 3 years	10.2	13.9	In the second grade of junior high school

Table 1 (continued)

Case	Sex	Age at surgery (years)	Date of operation	Pre-op symptoms and signs	Digital markings	Mean ICP (PCO_2)	Changes in symptoms and signs after surgery	Period of follow-up (years)	Age at present (years)	Status at present
11. A.S.	Boy	3.4	January 16, 2006	Few words Poor understanding	4	9 (38.1)	In primary school at age 6 Increased vocabulary, improved understanding, and ability to complete toilet training after 5 months 3-word sentences and social play after 2 years In the SE class of kindergarden	10.1	13.5	No problems in daily life In the second grade of SE junior high school Some difficulties in conversation and relationships with others
12. E.K.	Boy	1.6	February 8, 2007	Apparently normal development	3	13 (44.7)	Significant improvements in speech at 3 months In primary school at age 6	9.1	10.6	In fourth grade in a primary school
13. H.R.	Girl	7.5	April 19, 2007	Unable to use sentences, only words Difficulties in relationships with other children Hyperactive	4	13 (37)	–	8.8	16.3	In second grade of SE high school
14. G.Y.	Boy	7.9	June 14, 2007	Incorrect use of words Temper tantrums Head banging Hyperactive No recognition of danger Difficulty playing with other children	4	37 (42.2)	No behavior problems in school Greatly reduced head banging No problems with conversation, elimination of hyperactivity, and expression of empathy and healthy play with other children after 2 years	8.8	16.7	Unknown
15. G.R.	Boy	3.8	July 17, 2008	Normal development Headaches and vomiting	4		Used long sentences and displayed compassion after 1 month Elimination of headache and vomiting In primary school at age 6	7.6	11.4	In sixth grade of the primary school Well-adjusted socially No problems in daily activity
16. K.K.	Boy	5.6	September 25, 2008	Delayed language; <3-word sentences Poor pronunciation Difficulties in understanding play rules	4	11 (39.2)	Better conversation and pronunciation after 1 year In the SE class of primary school Good at arithmetic	7.4	13	In the SE class of junior high school
17. K.Y.	Boy	4.4	December 18, 2008	2-word sentences with poor pronunciation Difficulties in understanding Hyperactive Motor control difficulties	4	9 (36.2)	Improvements in motor control, vocabulary, and hyperactivity after 6 months In primary school at age 6; no problems in conversation and increased exercise capability.	7.2	11.6	In sixth grade of the SE class in primary school IQ 75 at age 11
18. O.Y.	Boy	5.1	September 3, 2009	No meaningful words Head banging Stereotyped movement; keeping rounding himself	4	11 (43)	Increasing language acquisition increased and head banging stopped after 2 months Stereotyped movement gradually decreased and communication improved after 2 years	6.6	11.7	In SE school Some difficulties in conversation No other problems in daily life activities Asymptomatic after surgery
19. H.R.	Boy	1.7	August 12, 2010	Seemingly normal development	4	16 (41.7)	Normal development	5.6	7.3	In primary school
20. S.K.	Boy	2	December 16, 2010	Normal development	3	13 (36.1)	Normal development	5.3	7.2	In primary school
21. I.K.	Boy	3.3	April 21, 2011	–	3	19 (35.5)	–	4.9	8.2	In primary school No regression Normal intelligence (IQ = 102) at age 8
22. N.S.	Boy	5.5	July 28, 2011	3-word sentences Difficulty making eye contact Hyperactive	3	8 (38.6)	–	4.6	10.1	In fifth grade SE class in primary school. Highly sociable; almost no problems in daily life
23. A.K.	Girl	5.8	November 1, 2012	Severe papilledema	4	13 (35.6)		3.4	9.2	In third grade SE class

Table 1 (continued)

	Case	Sex	Age at surgery (years)	Date of operation	Pre-op symptoms and signs	Digital markings	Mean ICP (PCO_2)	Changes in symptoms and signs after surgery	Period of follow-up (years)	Age at present (years)	Status at present
					Difficulties with conversation Hyperactive Unbalanced diet			After 1 month, had increasing vocabulary, improvement in conversation and understanding, cessation of hyperactivity and unbalanced diet Papilledema disappeared In SE class in primary school at age 6 At 1 year, no problems in conversation			Slightly impaired intelligence
	24. H.I.	Boy	3.9	March 27, 2014	Seemingly normal development	4	18 (37.7)	No hyperactivity and improved sense of balance	1.9	5.8	No developmental delays
	25. K.R.	Boy	2.3	October 16, 2014	No meaningful words Sitting with support Constant drooling Cortical thumbs	4	8 (41)	–	1.4	3.7	Speaking 2-word sentences Singing songs Running around
	26. S.A.	Boy	4	October 23, 2015	Mostly normal development; some language delay DQ = 86	4	19 (34.8)	Normal social play after 3 months DQ = 96 after 3 months	0.3	4.3	No problems in his daily life
	27. Y.S. 山村	Boy	7.9	January 15, 2016	Hyperactive Problems in comprehension of sentences	4	7 (40.6)	Cessation of hyperactivity immediately following discharge from the hospital At 1 month, expressed appropriate classroom behavior	0.1	8	No problems in his normal class
Type 2	28. O.I.	Boy	1.6	November 10, 1997	No meaningful words Hyperactivity Small (at least 2 standard deviations) head circumference	4	N/A	Reduced hyperactivity, ability to speak several words after 4 months After 14 months, able to make easy conversation Head circumference gradually increasing but still below normal In SE class in primary school After 4 years, conversed well At age 9, demonstrated intellectual impairment (IQ = 58)	18.3	19.9	In SE high school Trying to get a driver's license and job IQ (WISC4); 47 (May 30, 2016)
	29. N.R.	Boy	3.8	December 10, 1999	Speech delay; unable to use sentences Hyperactive DQ = 67 at age 44 months	0	N/A	Hyperactivity was reduced immediately after surgery At age 6, was speaking sentences; in regular primary school DQ = 88 Some difficulties in schoolwork in the second grade in regular primary school 6 years old 88 (February 12, 2012)	16.2	20	Graduated SE high school; working and obtaining driver's license
	30. A.K.	Boy	3.9	November 9, 2000	Limited to several words	2	10 (35.9)	After 1 month, could look at picture books After 2 months, vocabulary increased	15.4	19.3	Graduated from SE high school Simple conversation Working in a disability frame
	31. A.S.	Boy	2.5	August, 6, 2001	No meaningful words at age 28 months Hyperactivity No eye contact No response when calling his name	4	8 (43.6)	No improvement	14.6	17.1	Severe impairments of intelligence In SE high school
	32. K.Y.	Boy	4.8	October 6, 2003	Simple 2-word sentences Difficulties in conversation Hyperactivity Incomplete toilet training Head banging	4	12 (46.7)	–	12.4	17.2	In third grade of agricultural high school
	33. M.H.	Boy	3.5	November 1, 2007	Unable to walk at age 3; crawling No meaningful words	3	17 (42.2)	After 6 months, sleep respiration was rescued and drooling was markedly decreased	8.4	11.8	In the sixth grade of SE school; walking without any support

Table 1 (continued)

Case	Sex	Age at surgery (years)	Date of operation	Pre-op symptoms and signs	Digital markings	Mean ICP (PCO_2)	Changes in symptoms and signs after surgery	Period of follow-up (years)	Age at present (years)	Status at present
				Sleep apnea Constant drooling Often hospitalized due to respiratory problems			Able to stand at 1 year Able to walk with support at 2 years			No meaningful words; communicates using finger signs and gestures
34. K.K.	Boy	3.6	January 8, 2009	2-word sentences Difficulties with conversation Hyperactivity Temper tantrums Poor understanding	4	7 (39.3)	After 3 months, used 3-word sentences; temper tantrums and hyperactivity improved	7.2	10.8	In the SE class in primary school No problems in daily conversation Continued improvement in temper tantrums and hyperactivity
35. T.T.	Boy	8.4	October 3, 2014	From age 5, expressed head banging and paranoia From age 6, also reported headache and vomiting in the morning Hypotonic Difficulty forming new memories Additional cognitive regression	4	N/A	–	1.5	9.9	Keeping the normal class Remained some autistic tendencies

Digital markings are the area(s) of the skull found to have signs of damage during imaging, categorized as follows: 0, no markings; 1, occipital; 2, occipital and temporal; 3, occipital, temporal, and parietal or frontal; and 4, occipital, temporal, parietal, and frontal

SE special education

sphenoid bone on both sides were partially removed, after which the sphenoid ridge was excised up to the meningo-orbital band. In most patients, the sphenoid ridge was very thin and wide and entered between the frontal and temporal lobes. Finally, the supraorbital bar with orbital roofs was removed as one piece. The extracted frontal calvarium was cut into four parts and returned to the forehead after trimming. The orbital bar was returned and fixed outside the orbit with 4–0 nylon suture.

Posterior A linear craniectomy was performed by connecting four burr holes (bilateral parasagittal burr holes and bilateral burr holes made in the outermost part of the parietal bone) that were several centimeters posterior to lambda. The craniotomy was performed on both sides of the parietal bone, leaving the median bone of the sagittal suture. The extracted parietal bone was cut anteroposteriorly into three parts, and one part was returned. Three fracture lines were introduced in each side of the temporal bone. The returned parietal bone was allowed to float (Fig. 2).

The width of craniectomy approximately was 1–3 cm at the anterior part and 3–4 cm at the posterior part in patients under 4 years old. In 5 years of age or older, its width was narrower.

Results

Clinical assessment

Table 1 provides the clinical features of the patients and their outcomes (if known). Speech delay of various degrees was observed in 27 patients and described as follows: no meaningful word (n = 7), 20 words or fewer (n = 7), 100 words or more (n = 1), speaking in two-word sentences (n = 8), and speaking in three-word sentences (n = 4). Hyperactivity was observed in 15 patients and included excessive locomotion, inability to sit still, restlessness, and poor concentration. In 10 patients, autistic behavioral tendencies were observed, including reluctance to make eye contact, communication problems with other children, and spinning or other stereotypical movements. Seven patients had impaired motor function, primarily identified as being unable to walk by oneself or poor balance. Self-mutilation (mainly head banging) was observed in seven patients. Temper tantrums or panic were observed in eight patients. For example, one patient panicked and cried for over 30 min. Two patients presented with repetitive headache and vomiting. Cognitive regression was found in three patients, for whom previously spoken words or cognitive behaviors were lost over time.

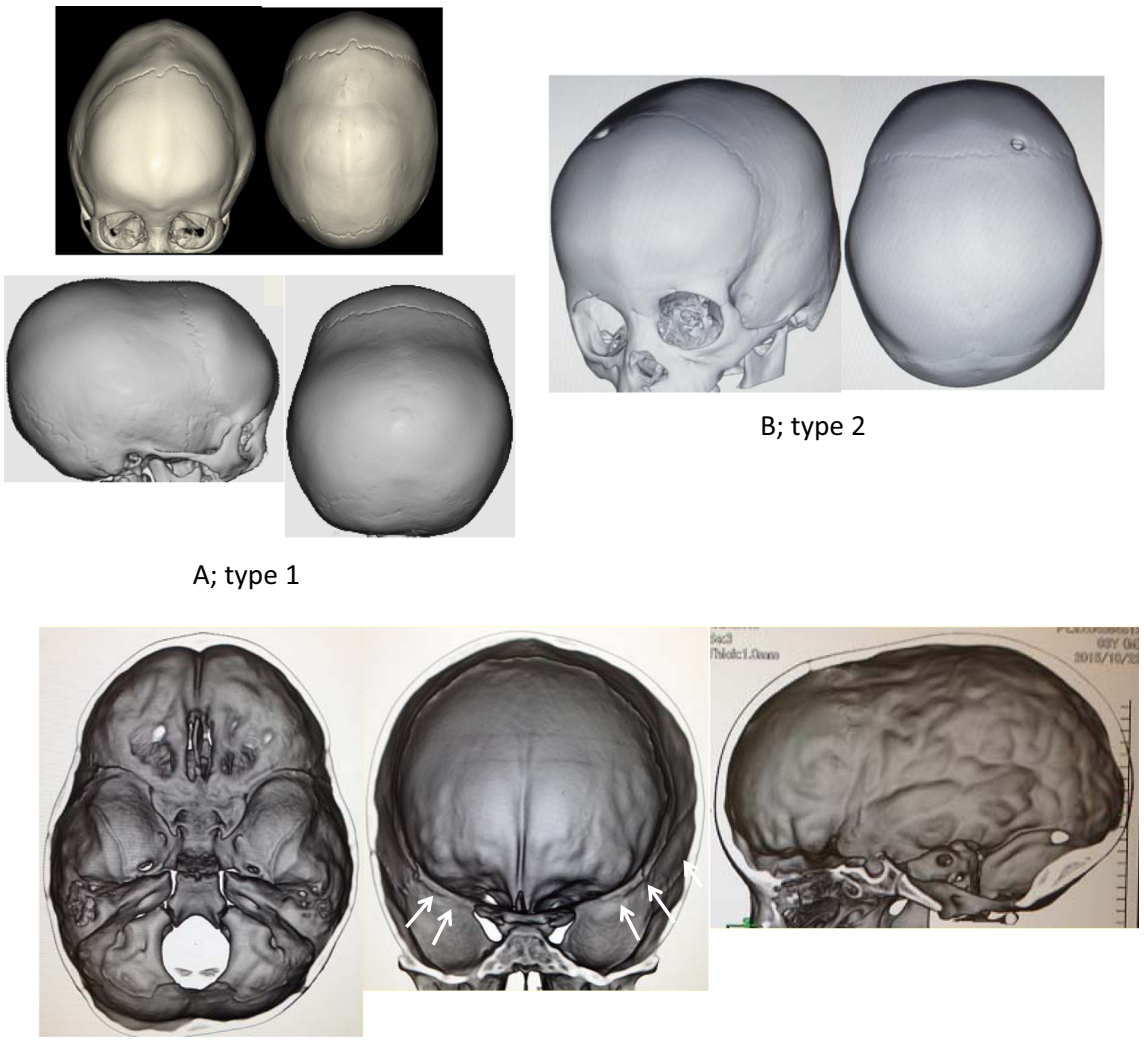


Fig. 3 Two types of metopic-sagittal synostosis. **a** Type 1, scaphocephalic type. *Upper column*: with the sagittal ridge (eight cases). *Lower column*: without the ridge (19 cases). **b** Type 2, non-scaphocephalic (eight cases total; no sagittal ridges evident). A metopic ridge was confirmed in all patients. **c** The smaller anterior fossa, the very thin and wider sphenoid ridge (*arrows*), and marked digital markings

Diagnostic assessments

Twenty-seven patients were diagnosed with scaphocephaly by visual inspection (type 1); in eight of those patients, a sagittal ridge was observed by palpation. Eight patients were normocephalic (type 2) and lacked a sagittal ridge. A metopic ridge was observed by visual inspection or palpation in all 35 patients, as well as mild frontal bossing. Hollow temples were also noted in all patients (Fig. 3).

Definitive diagnoses were performed by 3D-CT scans, in which the fusion of the metopic ridge and sagittal suture was confirmed in all patients. The sagittal ridges observed by palpation were also shown by a 3D-CT scan. Intracranial 3D-CT imaging revealed narrowing of the anterior cranial fossa in almost all patients. The imaging of the internal table of the calvaria revealed digital impressions in the whole skull in 26 patients and in the parietal area or reaching up to the forehead in six patients (Table 1). With MRI, we found one patient whose cerebellar tonsil extended to the spinal canal; no other brain malformations or abnormalities were found.

We have recently attempted imaging the sphenoid ridge. The ridge is obviously thin, and the ability to observe it varies widely between patients (Fig. 3). In one case, we found that although the sagittal suture was evidently present at the age of 8 months, closure mostly occurred at the age of 1 year and 3 months, with complete closure at 1 year and 8 months (Fig. 4).

Measurement of ICP

The mean values of PCO_2 and ICP for the first measurement were 31.2 and 8.3 mmHg, respectively, and those for the second were 39.2 and 19 mmHg, respectively. The second ICP

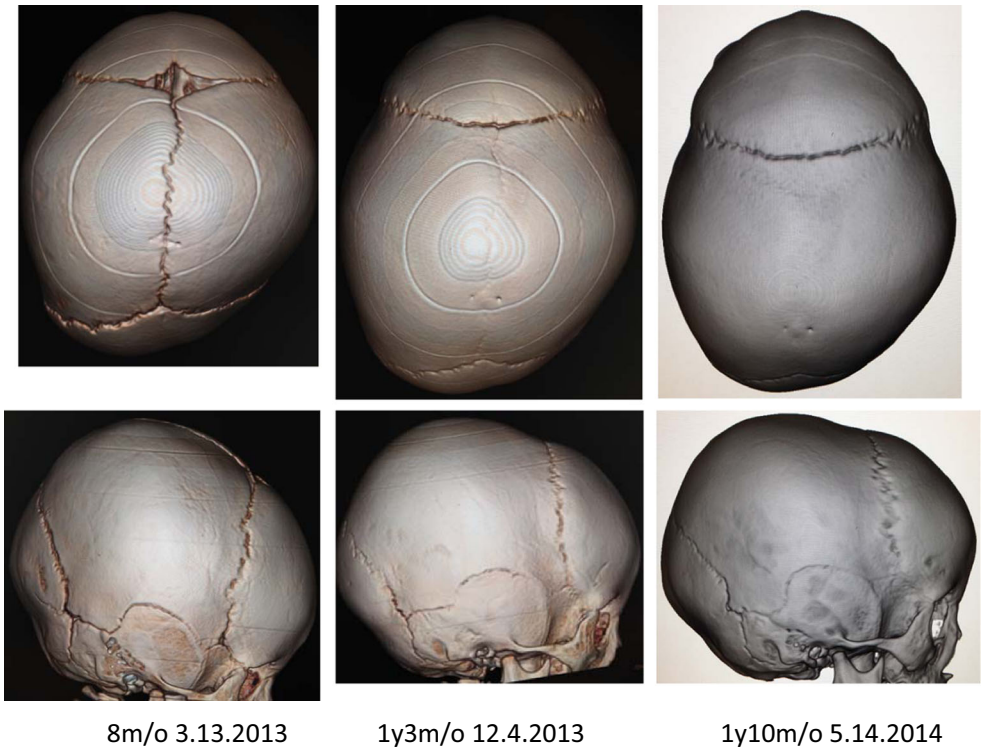


Fig. 4 The gradually closed sagittal suture

measurements ranged from ≤10 mmHg (n = 1), 11–15 mmHg (n = 4), to ≥16 mmHg (n = 30) and are given in Table 1). The ICP values were high in the patients having digital impressions over 75% or more of the skull, except for one patient (4 mmHg). However, ICP values were also high in two patients whose digital impressions were limited only to the occipital and temporal lobes.

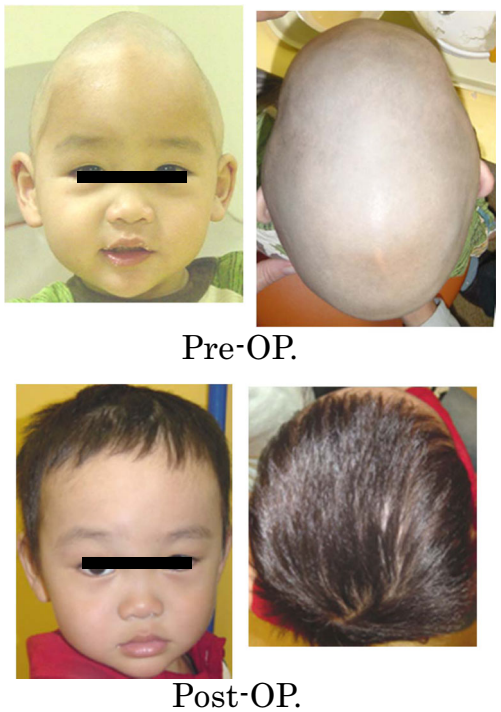


Fig. 5 Cosmetic changes in a type 1 case. *Upper column*: Images demonstrating the narrow forehead and scaphocephaly. The metopic and sagittal ridges are visible prior to surgery. *Lower column*: Images showing the wider forehead and normocephalic head shape after surgery

Results of the surgical procedure

No complications were observed in the 35 patients. Cosmetically, the forehead was widened and the frontal bossing eliminated in all patients. The degree of the hollow in the temples was also improved. All of the patients with type 1 scaphocephaly were normocephalic after surgery (Fig. 5).

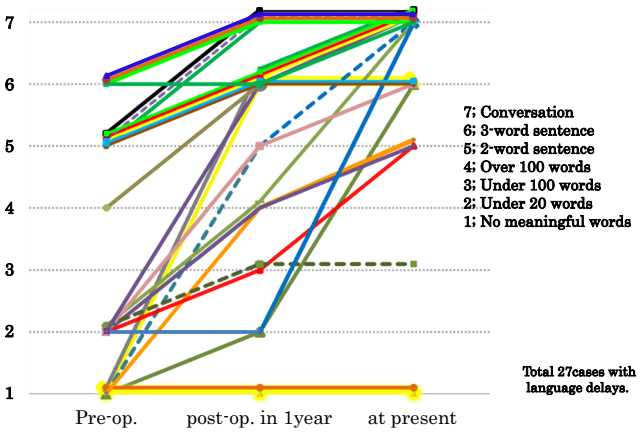
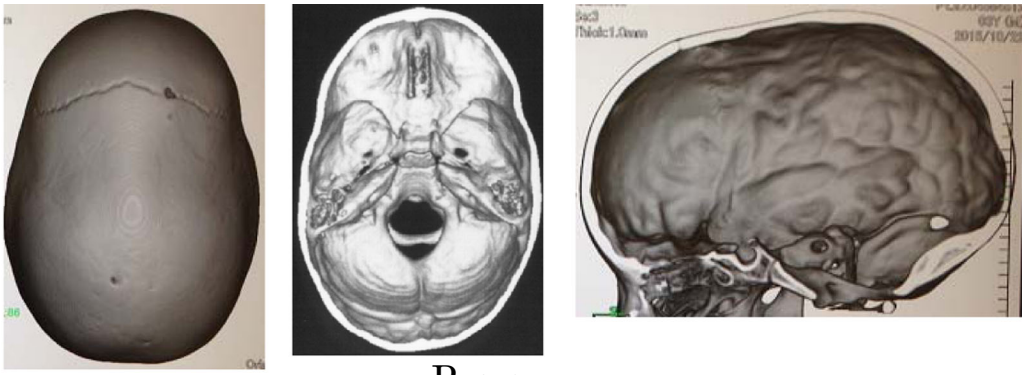


Fig. 6 Changes in speech delay, language, and acquisition following surgery



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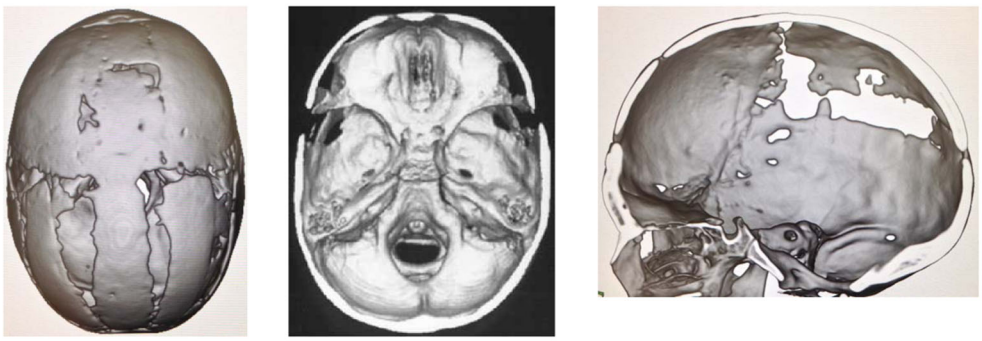
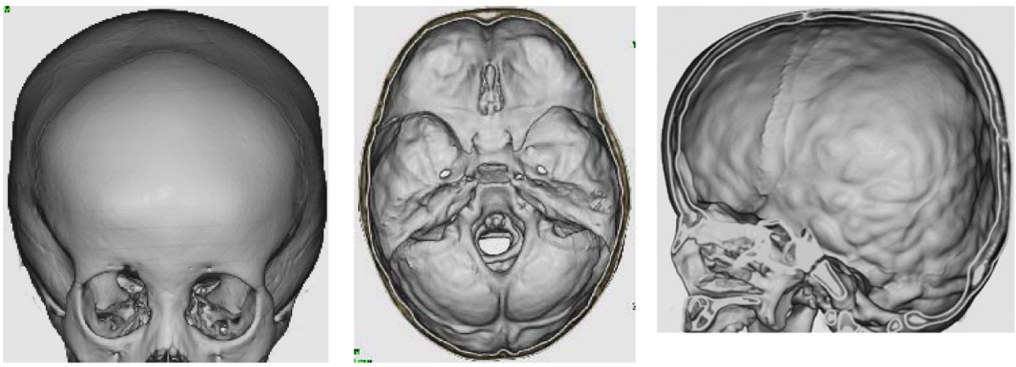


Post-op. 6months

A



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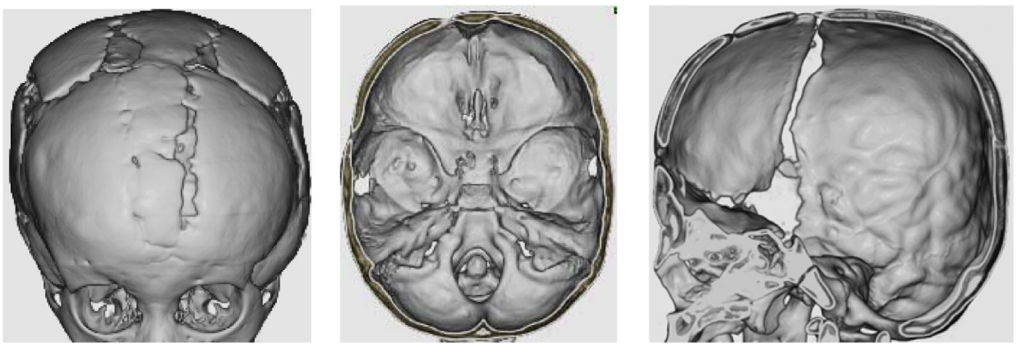

Post-op. 6months

B

◀ **Fig. 7** Changes induced by surgical intervention as identified by 3D computed tomography. In both **a** and **b**, the *top row* of images demonstrates the pre-operative condition, and the *bottom row* demonstrates the post-operative condition 6 months after surgery. **a** Scaphocephalic type (type 1). Note the enlargement of the frontal area and the anterior fossa, decrease of digital markings, and more normocephalic shape in the post-operative images. **b** Normocephalic type (type 2). Note the enlargement of the frontal area and the anterior fossa and decrease of digital markings in the post-operative images

Since bone regeneration was so fast, in 2 years post-op. bone defect almost closed in all patients.

Clinical outcomes

As seen in Table 1, speech delays improved in 25 of 27 patients. Of seven patients who used no meaningful words before surgery, two patients have not shown any change as of the most recent follow-up, but one and two patients improved so as to speak in two- and three-word sentences, respectively, and two other patients were able to have daily conversations without difficulty. In the seven patients who spoke ≤20 words, improvements included a slight increase in the number of words ($n = 1$), speaking in two-word sentences ($n = 2$), speaking in three-word sentences ($n = 1$), and no difficulty having ordinary conversations ($n = 3$). One patient who initially spoke ≥100 words was subsequently able to have ordinary conversation without difficulty. Of the eight patients who had spoken in two-word sentences before surgery, two patients improved to speaking in three-word sentences and six patients became able to have ordinary conversations. All four patients who had spoken in three-word sentences or more became able to have ordinary conversations (Fig. 6).

Improvements were observed in all 15 patients with hyperactivity, 8 of 10 patients with autistic tendencies, all 7 patients with motor impairments, all 7 patients with self-mutilation behaviors, and all 8 patients with panic/temper tantrum symptoms. In the two patients having repetitive headache and vomiting, these symptoms improved immediately after surgery. Moreover, surgery improved a sleep disorder in one patient, enabling the reduction of the patient's medicine for the disorder. Five patients whose development had been considered normal before surgery but who may have had minor speech delays and hyperactivity showed an increase in word expression and decrease in hyperactivity.

Improvements in clinical symptoms became evident within 3 months after surgery in most patients. There was no change in one patient who had undergone surgery at the age of 2 years and 4 months. This patient had speech delay (no meaningful words), high levels of hyperactivity, and autism-like symptoms before surgery. We were unable to contact two patients. Those patients whose symptoms improved soon after surgery have continued to thrive (Table 1).

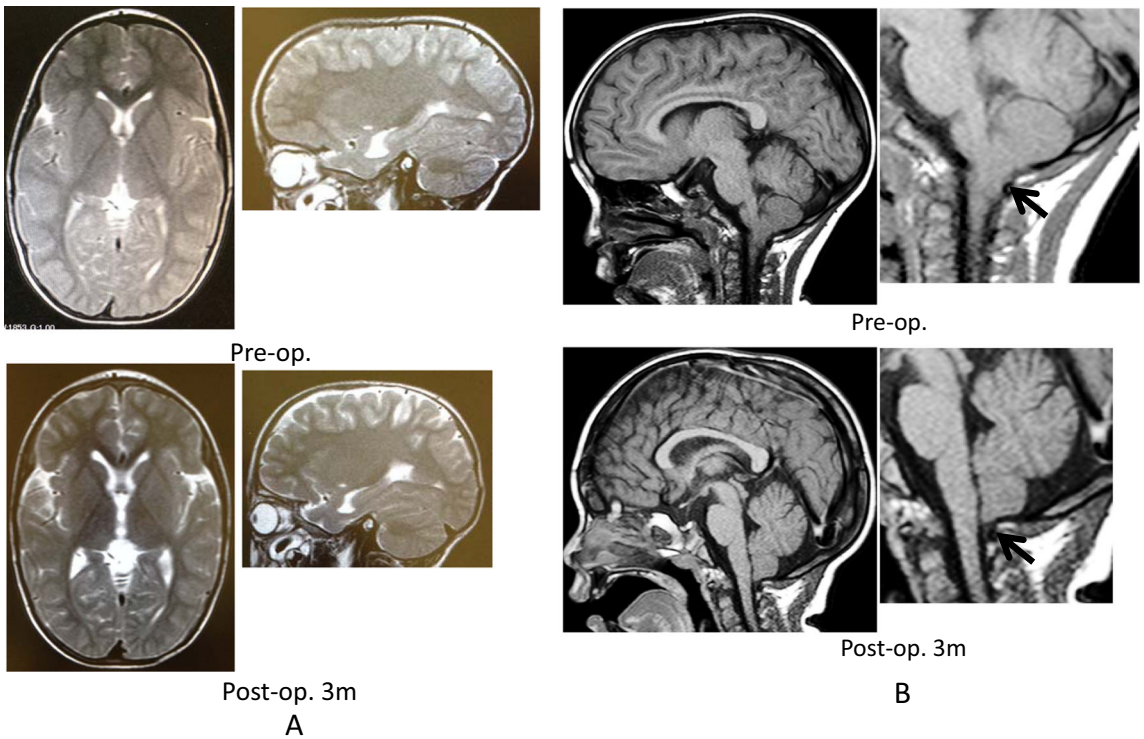

Fig. 8 Changes after surgery identified by MRI. **a** Note the scaphocephalic shape and smaller frontal lobes in prior to surgery. The normocephalic shape allowed for bigger frontal lobes, as seen in the post-operative MRI. **b** Note the finding of a tonsillar herniation (*arrow*) in this case prior to surgery (*upper column*), and the herniation completely disappeared after surgery (*lower column*)

In all patients, 3D-CT scans revealed the enlargement of the forehead and anterior cranial fossa. The type 1 patients (those with scaphocephaly) became normocephalic after surgery (Fig. 7). Scans demonstrated a decrease in digital impressions in all patients at 6 months after surgery, though this was not quantified. Bone regeneration allowed patients to experience daily life without difficulty by 1 to 1.5 years after surgery. MRI also showed the enlargement of the frontal lobe and the change was from the scaphocephalic to normocephalic skull (Fig. 8). Interestingly, the tonsillar herniation was observed before surgery in one patient disappeared after surgery (Fig. 8).

Discussion

After cranioplasty in a child patient with mild trigonocephaly, language delay, and hyperactivity led to symptomatic improvement in 1994, we have reported that a number of clinical symptoms in patients with mild trigonocephaly improve with surgery [20–23]. Notably, modern imaging has increased our identification of closure of the metopic and sagittal sutures in suspected mild trigonocephaly, as well as the fused metopic suture in patients with suspected scaphocephaly. This pathology is described as metopic-sagittal synostosis, which is recognized as a rare disease [3, 7]. Chumas et al. [3] only reported seven cases as previously unclassified cases of craniosynostosis. In our cases, all patients were diagnosed at the age of 1 year and older rather than in infancy. Although the patients who were diagnosed with scaphocephaly had a particular head shape that could have been diagnosed in infancy, the diagnosis was delayed long enough for most patients to exhibit symptoms. Because of these conditions, the degree of recognition concerning general craniosynostosis may suffer.

We made definitive diagnoses after observing a ridge of the metopic suture, as well as the fused sagittal suture, with 3D-CT scans. The sagittal suture usually does not form a ridge in most patients who have a normocephalic or scaphocephalic shape; this fact presumably makes diagnosis without neuroimaging difficult. However, the condition can be easily diagnosed via 3D-CT scan, which is additionally considered the best diagnostic tool to capture basilar changes as well [5].

It is thought that despite the closure of the sagittal suture (see Fig. 3), which would potentially impart a scaphocephalic head shape, the gradual fusion of the sagittal suture after birth allows for development of a normocephalic head shape in some of these patients [16].

We feel that the findings of the forehead are consistent with mild trigonocephaly in all 35 patients; however, the narrowing of the forehead and anterior cranial fossa and the wide sphenoid ridge were considered to be abnormal findings.

In 32 patients, 3D-CT scans showed digital impressions in the area of ≥3/4 of the skull. In the cases with more digital impressions, the measured mean ICP value was high (≥10 mmHg). These results suggest the two phenomena may be positively correlated. In fact, there was a decrease in digital impressions after surgery, and in many of the patients for whom the impressions were decreased, ICP was also reduced. The tonsillar herniation in one patient, which was eliminated by the surgery, was further evidence of high ICP in at least some members of the group; it too was eliminated by the surgery. Thompson et al. [26] reported that overnight subdural intracranial pressure monitoring showed an increase in ICP relatively frequently in sagittal and metopic suture synostosis. Since most of their patients were ≤2 years old, and most of our patients were ≥2 years old, ICP may tend to increase as patients develop.

Concerning the surgical procedure for the anterior part, we used a procedure that had been performed for mild trigonocephaly, in which the upper orbit was extracted as a bar and shifted slightly anteriorward, the sphenoid bone was removed, and the abnormally broad sphenoid ridge was then also sufficiently removed. Some earlier studies pointed out the importance of an approach to the anterior cranial fossa in brachycephaly and plagiocephaly [1, 6, 13, 14, 17, 19], and we have followed this procedure. We performed osteotomy in the coronal section, allowing for a sufficient reduction in pressure. Fast bone regeneration is an interesting component of this pathological condition [13]. Indeed, we have had phenomena where bone regeneration occurred very early, even if a large osteotomy was performed in infancy. This phenomenon may be a cause of the recurrence of the pathology.

We selected this procedure in order to achieve a sufficient decrease in intracranial pressure and prevent the postoperative shape from regressing to the original shape. In fact, the patients with scaphocephaly became normocephalic after surgery, had no cosmetic problems, and had markedly reduced digital impressions suggestive of a decrease in intracranial pressure. Bone regeneration by 1 to 1.5 years after surgery had progressed well enough for patients to live their daily lives without difficulty.

Clinical symptoms had developed in 30 patients, including language delay, hyperactivity, autistic tendencies, motor impairment, self-mutilation, and panic/temper tantrum; all symptoms improved at high rates after surgery. In the patients showing improvement, some symptoms started to improve immediately after surgery and steadily improved thereafter. The enlargement of the forehead and anterior cranial fossa due to surgery caused the enlargement of the volume of the frontal lobe, which is considered to contribute to the reduction in symptoms. Moreover, an abnormally broad sphenoid ridge, which had been observed in our previous cases of mild trigonocephaly, was also seen in the patients of the present study. McCarthy et al. [14, 15] described this abnormal sphenoid ridge as a heavily overgrown sphenoid ridge and recommended excising it. Because decreased blood flow in the

operculum has been associated with autism spectrum disorders in children [4], we believe that the reduction in autistic tendencies observed in our patients was caused by the decompression created by the removal of the sphenoid ridge contacting the operculum [23].

Since headache and vomiting, symptoms of high ICP, disappeared immediately after surgery, the elevated intracranial pressure was apparently improved after the decompressive cranioplasty. We believe the sleep disorder noted in one patient was also rescued by the surgical relief of the patient's high ICP, given the published evidence of abnormally high ICP during sleep in cases of craniosynostosis [18, 26]. Faster development after surgery in the five seemingly normally developing patients (all scaphocephalic) was also presumably caused by the improvement in the poor intracranial environment.

Neurodevelopmental problems in various types of craniosynostosis have been mainly described by psychologists [2, 9, 11, 24, 25, 27]. Although the patients in the present study had at most minor changes in brain morphology, the narrowed forehead and elevated intracranial pressure are considered to have negatively affected the brain, thus producing the symptoms. Since Kapp-Simon et al. [10] indicated that even minor brain deformity can cause clinical symptoms after a long period of time; such symptoms in our cases would be expected.

Considering the above descriptions, definitive surgical indications for metopic and sagittal synostosis diagnosed based on 3D-CT scans should include any symptoms produced by synostosis-induced, long-standing morphological changes and the increased intracranial pressure suggested by the marked digital impressions.

Aside from our reports [20–23] and a study by Inagaki [8], no studies have shown that symptoms were improved by adding a surgical procedure to the therapeutic course. Thus, the evidence of reducing symptoms surgically is scant [25]. However, we believe that metopic-sagittal synostosis may be more common than once thought, so further evidence in support of decompression surgery may be forthcoming. Neurosurgeons dealing with various craniosynostosis must know that patients with these pathologies might develop not only cognitive impairment but also behavioral problems such as hyperactivity and autistic tendency. In this report, psychological tests were not applied. In the future, to evaluate these symptoms, developmental tests must be applied, a language assay, the child behavior checklist, and the childhood autism rating scale. Finally, we designate this pathological condition for type 1 as trigono-scaphocephaly.

Compliance with ethical standards

Conflict of interest No conflict.

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