Child's Nerv Syst (2002) 18:215–224
DOI 10.1007/s00381-002-0568-1

ORIGINAL PAPER

Takeyoshi Shimoji
Satoshi Shimabukuro
Seiichi Sugama
Yasuo Ochiai

Mild trigonocephaly with clinical symptoms: analysis of surgical results in 65 patients

Received: 23 November 2001
Revised: 23 January 2002
Published online: 7 May 2002

T. Shimoji (✉)
Department of Neurosurgery,
Okinawa Naha Prefectural Hospital,
1-3-1 Yogi, Naha City,
Okinawa 902–0076, Japan
e-mail:
simoji_takeyosi@hosp.pref.okinawa.jp
Tel.: +81-98-8533111
Fax: +81-98-8323091

S. Shimabukuro
Department of Pediatrics,
Okinawa Naha Prefectural Hospital,
1-3-1 Yogi, Naha City,
Okinawa 902–0076, Japan

S. Sugama
Department of Pediatrics,
Tokyo Metropolitan Maternity
and Child Health Institute,
1–27–33 Sakura, Setagaya-ku,
Tokyo, Japan

Y. Ochiai
Department of Pediatrics,
Center for Development
of Children in Okinawa,
Awase, Okinawa City, Japan

Abstract *Introduction:* It has been believed that isolated, mild trigonocephaly rarely presents with clinical symptoms.
Patients and methods: We diagnosed and operated on 65 patients with mild trigonocephaly and developmental delay up to July 2000. There were 47 boys and 18 girls in our series. All patients had symptoms such as delay in language development, hyperactivity, autistic tendencies, and motor dysfunctions. Their facial features were characterized by a metopic ridge, depressed temples, heel-shaped rather than keel-shaped forehead, and slight hypotelorism. The most important physical sign was the palpable metopic ridge. Most patients did not exhibit any symptoms until they were more than 1 year old. Fifteen patients showed regression in language acquisition and use. Three-dimensional computed tomography revealed the metopic ridge, depressed pterional regions, hypotelorism, and small anterior fossae. Magnetic resonance imaging was performed on all patients and demonstrated no abnormal findings in the brain. Single-photon emission computed tomography (SPECT) was performed on 83% of patients and revealed decreased cerebral blood flow (CBF) in the frontal lobes of 76% of those patients. Decompressive cranioplasty of the frontal bone involving the skull base was performed on all patients. *Results:* In most (61 out of 65) patients a degree of postoperative improvement in clinical symptoms was noted, especially in behavioral problems. Postoperative SPECT demonstrated increased CBF in the frontal lobes in 95% of the patients.
Conclusion: Based on these results, it can be postulated that mild trigonocephaly is frequently associated with developmental delays and that these symptoms can be improved to a certain degree by decompressive cranioplasty.

Keywords Trigonocephaly · Cerebral blood flow · Developmental delay · Decompressive cranioplasty

Introduction

In 1962, Anderson et al. [2] proposed that trigonocephaly might cause mental defects due to constriction of the frontal lobes by the narrow anterior cranium. However, subsequent reports indicated that patients with trigonocephaly rarely manifested mental problems [8, 9, 10, 11, 12]. In 1996, Collmann et al. [6] reported that mental deficiencies are not evidence of frontal lobe constriction and stated that such deficiencies are a common feature of syndromic trigonocephaly, probably due to coincidental cerebral maldevelopment. Collmann et al. concluded that surgery for trigonocephaly is mainly performed for cosmetic reasons. However, some recent reports have described a high rate of developmental delay in patients with trigonocephaly [5, 15, 28].

We initially treated several patients with "mild trigonocephaly" who presented with clinical symptoms such as delay in language development, hyperactivity, autistic tendencies, and motor delay and who showed improvement after undergoing a decompressive surgical procedure [27]. After that experience, we began to believe that there was a relationship between the aforementioned symptoms and the presence of mild trigonocephaly. When a care facility for children with developmental delays that was attached to a medical institution became aware of our experience, we were overwhelmed by referrals of children with trigonocephaly. This paper reports on the associated clinical symptoms, neuroradiological findings, and surgical results in a series of 65 patients and stresses the relationship between the aforementioned symptoms and mild trigonocephaly.

Patients and methods

Patients

From October 1994 to June 2000, 65 children diagnosed with mild trigonocephaly, mainly in the previous 2 years, were examined in the Division of Neurosurgery, Okinawa Prefectural Naha Hospital, Naha, Japan. Two patients were infants less than 1 year of age, 2 were 1–2 years old, 6 were 2–3 years, 7 were 3–4 years, 15 were 4–5 years, 11 were 5–6 years, 10 were 6–7 years, 10 were 7–8 years, and 2 were 8–9 years of age. All the children were evaluated by pediatric neurologists. Surgery was performed on children less than 8 years of age; 47 of these were boys and 18 were girls. All the patients following patient 6 underwent chromosome testing, but no abnormal results were found. All 65 patients were considered to be in the nonsyndromic trigonocephaly category.

Fifteen patients (11 boys and 4 girls) with the same symptoms were diagnosed with mild trigonocephaly for which surgical treatment was indicated but their parents did not permit surgery. These patients received only rehabilitation in institutions, kindergartens, or schools and served as a control group for comparison with the patients we treated surgically.

Preoperative evaluation

Facial features

The patients' foreheads typically resembled a heel rather than the more usual keel shape. The metopic ridge was easily palpated but was not situated higher than in patients with typical trigonocephaly. The temples appeared to be depressed bilaterally. Hypotelorism was not remarkable physically, but was noted on X-ray (Fig. 1, top panels). Milder cases of trigonocephaly are difficult to diagnose on the basis of physical appearance alone because of the shallower metopic ridge and flatter forehead in those patients. In our patients' the diagnosis was therefore based on three-dimensional computed tomography (3D-CT) imaging.

Symptoms

Two interesting findings were noted preoperatively. First, most patients (58 of 65) were not diagnosed before they were 1 year old. Secondly, 15 of the 65 patients had exhibited regression in language acquisition and use. Other symptoms included global delays

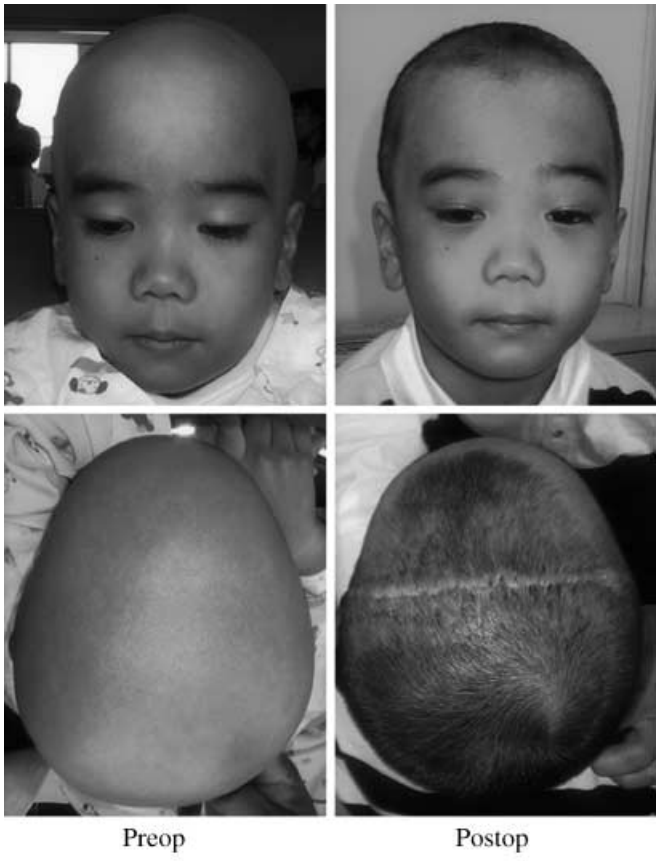

Fig. 1 Changes in a representative patient's facial features after surgery. *Left panels*, before surgery; *right panels*, after surgery. Note the metopic ridge, depressed temples, and narrow, heel-shaped forehead in the preoperative photograph. These features resolved postoperatively

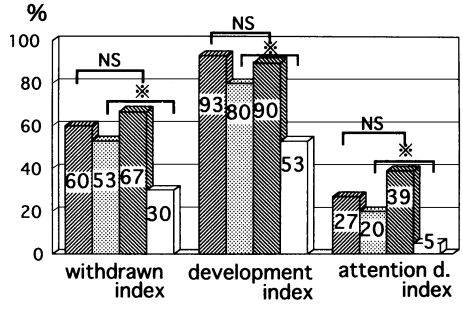

Fig. 2 Comparison of JCBCL index scores in the control (n=15) and surgical group (n=43) at baseline and at 6 months. The numbers in the *bars* represent mean scores for the respective groups. *Stars* the difference was statistically significant (P=0.05), *NS* not significant, *upward sloping lined bars* first (baseline) score in controls, *dotted bars* score for each index in the controls at 6 months, *downward sloping lined bars* first (baseline) score for each index in surgical patients, *white bars* score for each index in surgical patients 6 months after surgery

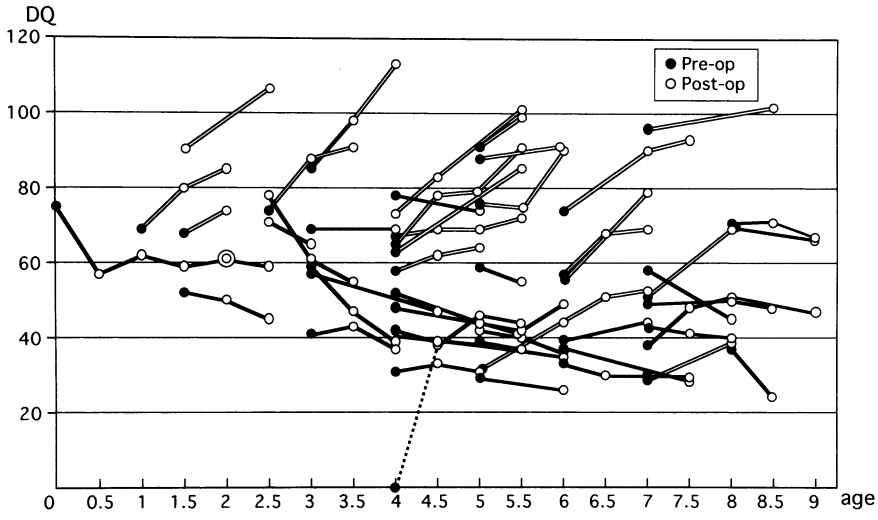

Fig. 3 Chronological changes in DQ in the surgical group before and after surgery. White lines, upward trend (20 patients [41%]); black lines, downward trend (29 patients [59%])

Table 1 Improvement in symptoms of mild trigonocephaly after surgery (*n*=65)

Symptom	Present before surgery (no. of patients)	Improved after surgery (no. of patients)
Language delay[a]	61	37
Hyperactivity	41	39
Impaired social interactions	30	23
Motor dysfunction	8	7
Self-mutilation	10	9
Panic and irritability	35	32

[a] No spoken vocabulary, only a few words, one-way conversation, etc.

Table 3 Changes in language comprehension in the control (*n*=15) and surgical groups (*n*=44) at baseline and after 6 months (comprehension: *A* good conversation, *B* obey orders but insufficient understanding, *C* obey simple orders but insufficient understanding, *D* unable to understand any questions)

Control group		Surgical group	
Baseline	After 6 months	Baseline	After 6 months
3B	3B	11B	8A, 3B
8C	2B, 6C	25C	10C, 15B
4D	1C, 3D	8D	7C, 1D

Table 2 Changes in language use and acquisition from before to after surgery (*n*=61)

Level of speech ability	Before	After						
		No speech	Few words	10+ words	Two-word sentences	Sentences of 3+ words		
						No conversation	Difficulty in conversation	Fluent conversation
No speech	18	12	3	2	–	–	–	1
Few words only	11	–	6	1	3	–	1	
More than 10 words	12	–	–	3	7	–		2
Two-word sentences	6	–	–	–	–	–	2	4
More than 3-word sentences	14 (no conversation)	–	–	–	–	2	3	9

in language development, behavioral problems (hyperactivity, inappropriate social interactions, self-mutilation, panic, and irritability), and motor dysfunction (Table 1). In 61 of the patients, language development was delayed at different stages, in forms ranging from an inability to utter meaningful words to being unable to sustain a conversation with others despite the ability to speak three-word sentences (Table 2). Patients with delays in language development also had difficulty in comprehending speech. Language difficulties in 44 patients (all consecutive patients following patient 18, as evaluated by S. Shimabukuro) were analyzed and compared with those in 15 mild trigonocephaly patients who did not undergo surgery, and the results are presented in Table 3.

Hyperactivity and/or attention deficit/hyperactivity disorder was noted in 41 of the 65 patients. The related symptoms included difficulty in sitting still, constant fidgeting with the hands or feet, wandering away from parents, and excessive running and climbing behavior. Thirty patients exhibited impaired social interactions, manifested by aloofness, difficulty in maintaining eye contact, and lack of social or emotional reciprocity. Self-mutilation behavior, mostly in the form of head banging, was noted in 10 pa-

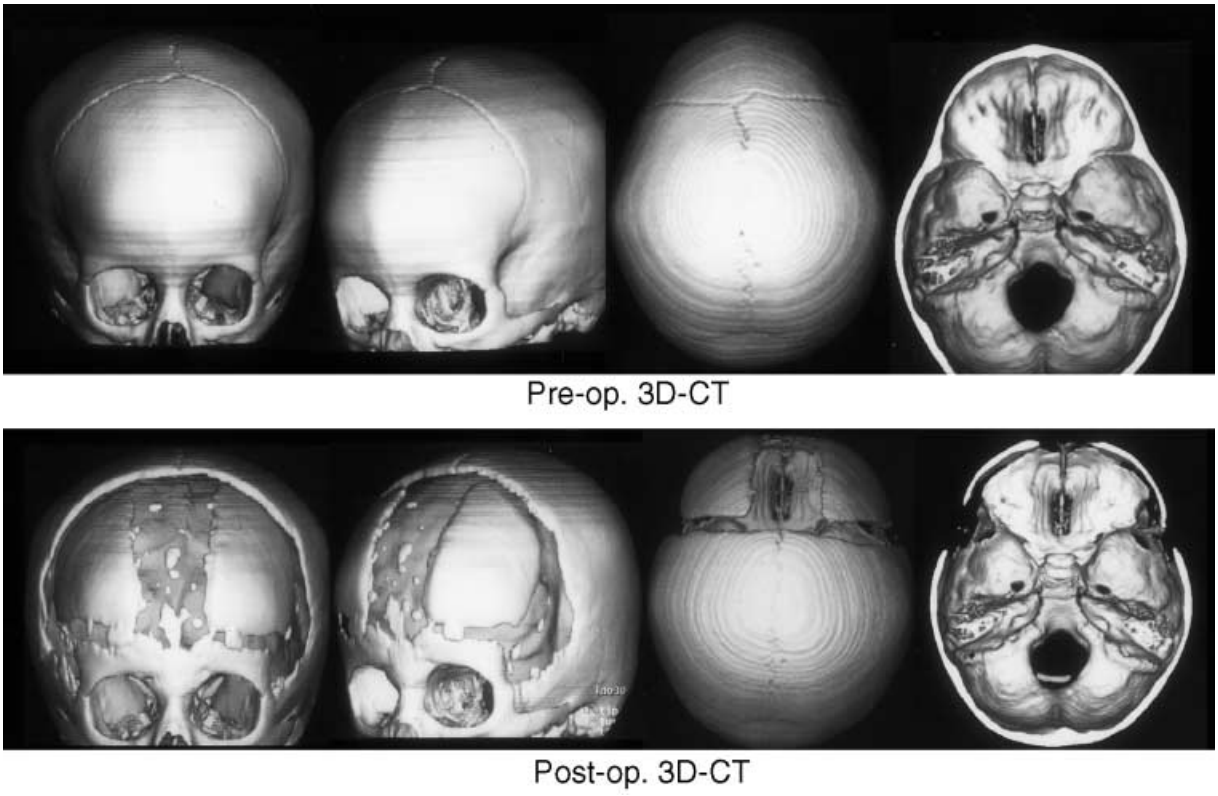

Fig. 4 Characteristic 3D-CT findings in patients with mild trigonocephaly. Preoperative 3D-CT (*upper panel*) clearly showed the metopic ridge, narrow, heel-shaped forehead, depressed temples, constricted anterior cranium, and hypotelorism. Three-dimensional CT performed 2 months postoperatively (*lower panel*) showed that the forehead had become wider and flatter and the anterior cranium had enlarged

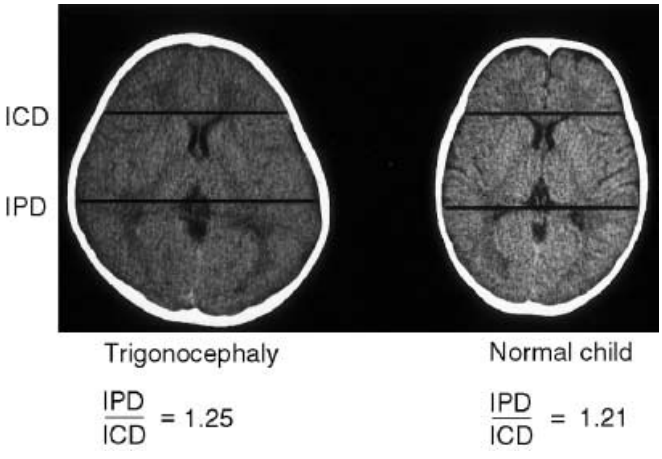

Fig. 5 Interparietal distance (*IPD*)/intercoronal distance (*ICD*) ratios in normal children (n=35) and mild trigonocephaly patients (n=50). The difference between the two groups was statistically significant (P=0.000 by Student's t-test)

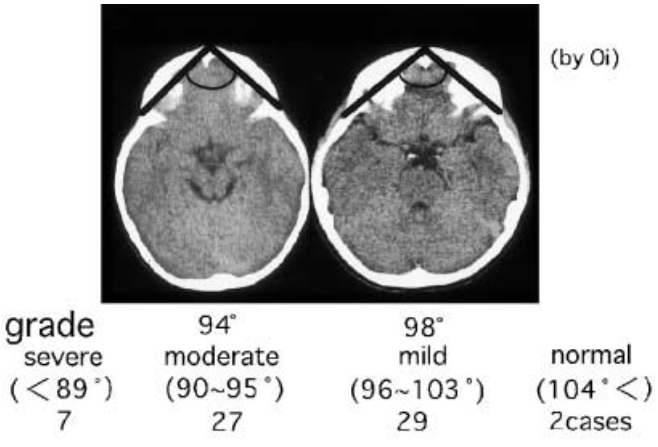

Fig. 6 Grade of trigonocephaly based on the nasion–pterion angle (Oi and Matsumoto's classification system [19, 20])

tients. Thirty-five patients exhibited panic and irritability when they were prevented from doing what they wanted. These episodes tended to last for quite some time.

The severity of behavioral problems in the 65 patients was analyzed using the Japan Child Behavioral Check List (JCBCL), which is based on Achenbach's 1992 profile [1]. The JCBCL consists of 100 questions divided into eight indexes by type of problem. Our 65 patients scored high on the three indexes for withdrawal, developmental delay, and attention deficit disorders. The results for each index in 43 patients are illustrated in Fig. 2 and compared with those obtained in the 15 nonsurgical controls.

Eight patients had motor dysfunction, manifested as not being able to sit, walk, jump, or ride a tricycle at the appropriate ages

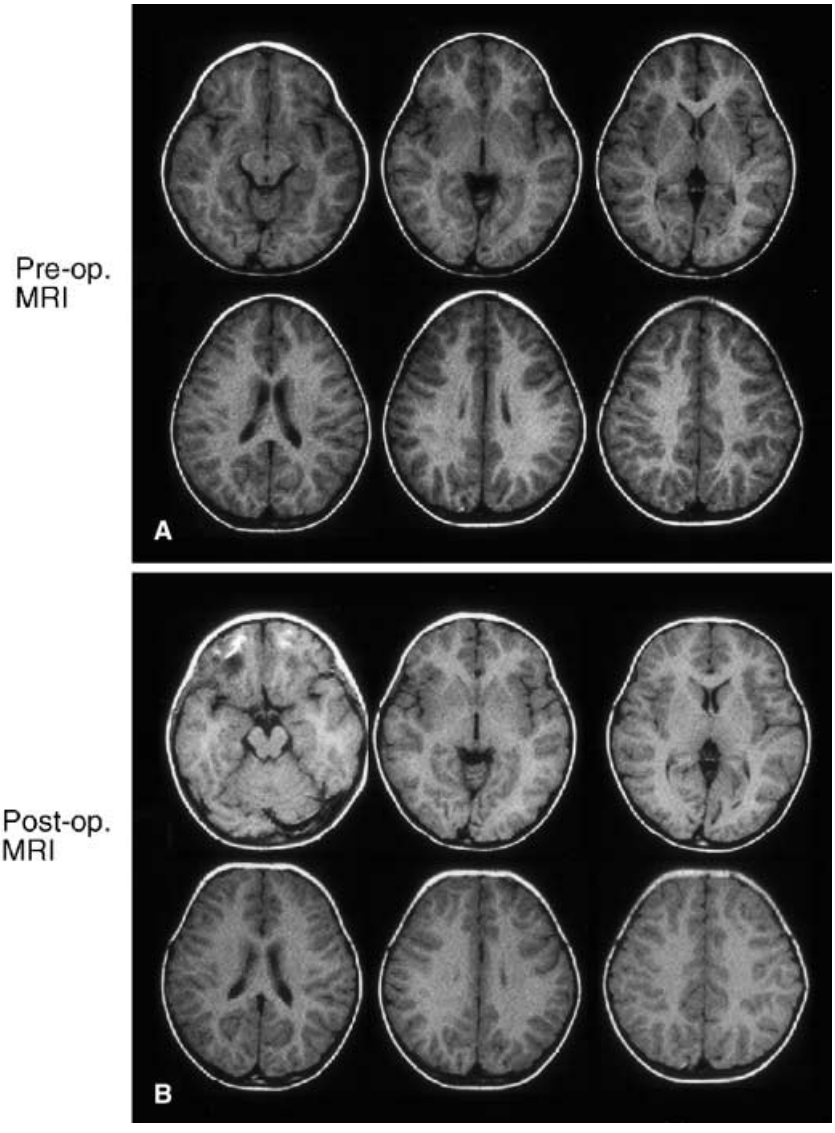

Fig. 7 Representative MRI findings **A** before and **B** after surgery in a trigonocephaly patient. Preoperative MRI showed constricted frontal lobes and no other abnormalities in the brain. Postoperative MRI showed that the constriction had disappeared and the frontal lobes had expanded markedly

and physical awkwardness, etc. The developmental quotient (D.Q.), as measured by the K-form Developmental Test (a commonly used test in Japan), was administered to all surgical patients pre- and postoperatively (Fig. 3).

Neuroradiological findings

Skull X-ray

Skull X-rays showed sclerotic changes of the metopic suture and mild hypotelorism. Since the anterior–posterior view was X-rayed in all patients, the interorbital distance (IOD) could not be compared with the standard IOD. The IODs on 3D-CT were measured in 65 patients with trigonocephaly (ranging in age from 1 to 8 years, mean 4 years) and in 28 healthy children (ranging in age from 1 to 7 years, mean 4 years). The mean IOD was 18.8 mm in the 65 trigonocephalic patients and 20.3 mm in the healthy controls. The difference was statistically significant (P=0.003, Student's t-test). In 61% of patients conspicuous digital markings were seen on the skull X-ray.

Computed tomography

All patients underwent helical CT scanning. Standard axial CT showed no abnormalities in the brain except for the small frontal lobe size. The characteristic 3D-CT findings of metopic ridge, small anterior fossae, depressed pterional regions, and hypotelorism were seen. The foreheads uniformly exhibited a heel-shaped deformity (Fig. 4).

Frontal stenosis was evaluated by calculating the ratio of the intercoronal distance (ICD) and the interparietal distance (IPD) according to the method reported by Bottero et al. [5]. The IPD/ICD ratio was calculated in 35 normal children who underwent CT after head injury and in 50 of our 65 patients. The ratio was 1.21 (SD 0.03) in normal children and 1.25 (SD 0.04) in the mild trigonocephaly patients. The difference was statistically significant (P=0.000, Student's t-test) (Fig. 5). Based on the CT imaging results of the nasion–pterion angle, the 65 trigonocephalic patients were divided into 7 severe cases, 27 moderate cases, and 29 mild cases, 2 being classified as normal, according to the method reported by Oi and Matsumoto [19] in 1986 (Fig. 6).

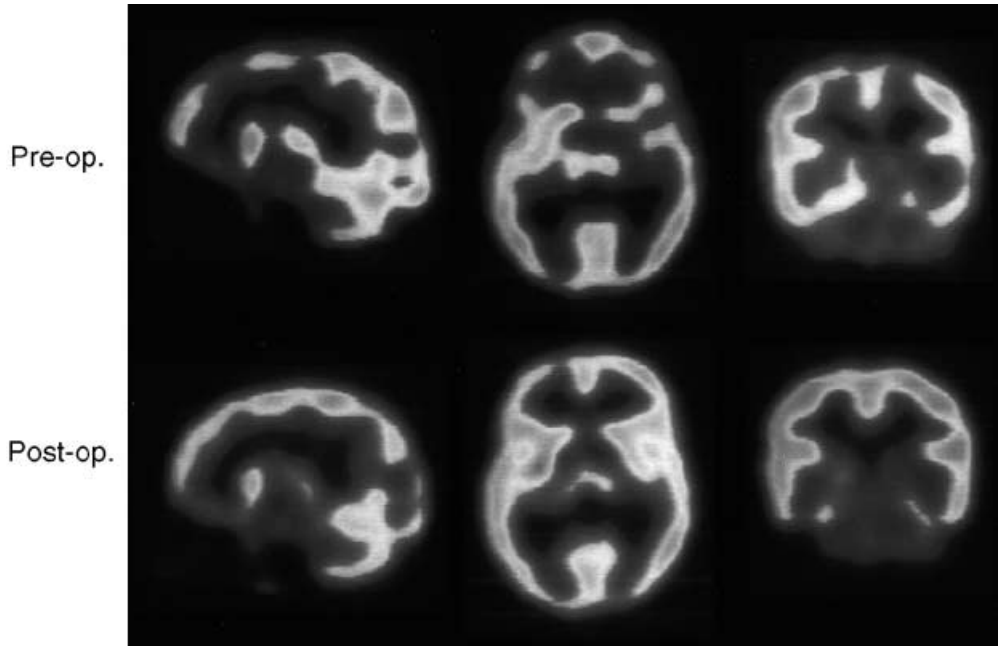

Fig. 8 Pre- (*upper panel*) and postoperative (*lower panel*) SPECT findings in sagittal (*left*), axial (*middle*), and coronal (*right*) slices. Preoperative SPECT showed decreased CBF in the upper and medial portion of the frontal lobes. Postoperative SPECT showed improved CBF in the same areas

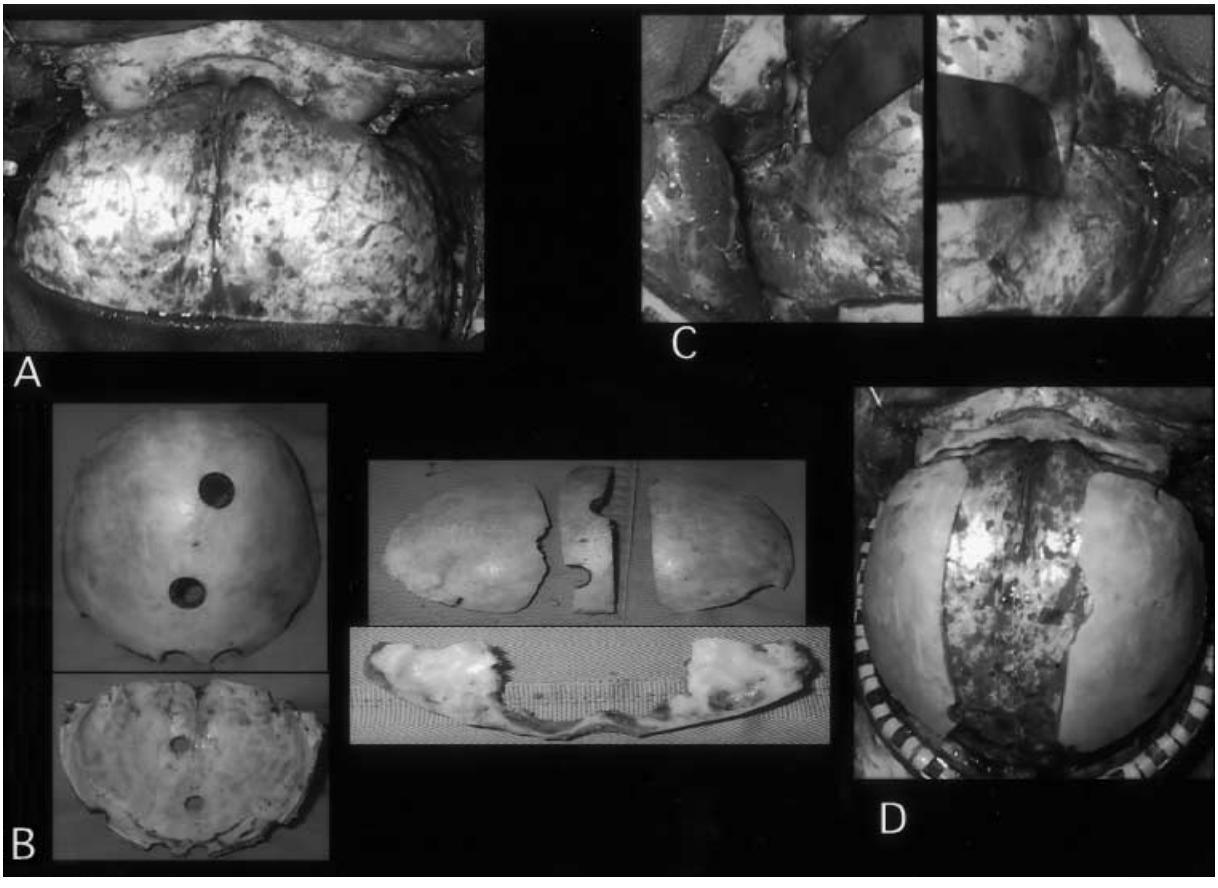

Fig. 9A–D Operative views. **A** After bilateral frontal craniotomy, behind the coronal sutures, the supraorbital bar with orbital roofs was removed. **B** Removal of the frontal bone. Note the digital markings on the calvarium. The frontal bone is divided into three pieces. **C** The greater and lesser wings of the sphenoid bone were removed with a rongeur. **D** The supraorbital bar was replaced with one stay suture at the side of the each lateral orbital rim. The trimmed frontal bones were replaced

Magnetic resonance imaging

Magnetic resonance imaging (MRI) was performed on all patients. As with CT, no abnormal findings were seen except for the reduced size of the frontal lobes (Fig. 7).

Single-photon emission computed tomography

Single-photon emission computed tomography (SPECT) with technetium-99m ethyl cysteinate dimer was performed on 54 of the 65 patients both pre- and postoperatively. Preoperatively, the results indicated that normal cerebral blood flow (CBF) was present in 13 and decreased CBF, mainly in the frontal lobes, in 41 patients (76%) (Fig. 8). These must be regarded as qualified results, however, since there is no standard for SPECT evaluation of CBF in childhood.

Surgical procedure

A bifrontal craniotomy was performed on the patients undergoing surgery, with burr holes situated bilaterally under the most anterior extension of the superior temporal line just above the frontozygomatic suture ("keyhole procedure") at the point posterior to the pterions at the paramedian points 2 cm behind the coronal sutures and over the glabella. Two burr holes on the metopic ridge made it easier to separate the superior sagittal sinus from the frontal bone. The frontal calvarium was removed in one piece. The outer third of the lesser and most of the greater wings of the sphenoid bone were bilaterally removed with a rongeur. The supraorbital rims were removed as a bar by dissecting through the frontozygomatic sutures, lateral rims, and roof of the orbits and through the bottom of the frontal bone at the midline. When the frontal sinuses were opened in older patients (more than 6 years of age), they were completely obliterated using pieces of temporal muscles and covered with periosteum. The orbital bar was reapproximated and loosely moored in position with only one suture at each side of the lateral rims of the orbit. Then the frontal calvarium was divided into three pieces (i.e., two frontal flaps and the metopic ridge). The frontal flaps were trimmed to size and replaced on the frontal lobes in a floating fashion (Fig. 9). The operative procedure described above was performed on all patients.

Results

Facial features

Except in the most severe cases, the patients did not have serious cosmetic problems preoperatively. However, their facial features were altered slightly postoperatively, with flatter and wider foreheads, flatter temples, and slightly wider spaces between their eyes (Fig. 1, top panels).

Symptoms

Of the 61 patients with delay in language development, 37 exhibited postoperative improvement (Table 1). However, patients who used fewer meaningful words prior to surgery showed less improvement, and those with less severe language problems tended to be able to engage more smoothly in conversations postoperatively. Table 2 shows the differences in the use of language after surgery, with patients divided into four groups and compared with the nonsurgical controls. Patients' comprehension of speech also improved after surgery. For example, most of those who were not able to obey a simple command such as "don't do that" before surgery were able to comprehend and obey a few months afterwards (Table 3).

Thirty-nine of the 41 patients with preoperative hyperactivity had improved to some degree 1–2 months postoperatively (Table 1); for example they stopped wandering away from their parents in shopping centers and department stores. The reduced hyperactivity allowed some to go with their families to fast-food restaurants for the first time, and they developed the ability to sit still for longer periods. Postoperative improvement was also noted in 23 of the 30 patients who exhibited impaired social interactions. For example, the children became more interested in their surroundings, were able to maintain eye contact, listened to others, smiled more often, expressed themselves more pleasantly, and showed greater emotional reciprocity within a few months after surgery (Table 1).

When behavioral problems were evaluated preoperatively and 6 months after surgery using the JCBCL, the preoperative withdrawal index scores of 60% in 15 controls and 67% in 43 surgical patients had decreased to 53% and 30%, respectively, at the later evaluation. The developmental index was 93% in the 15 controls and 90% in the 43 surgical patients before surgery, but 80% and 53%, respectively, by 6 months after surgery. Similarly, the attention deficit index was 27% in the 15 controls and 39% in the 43 surgical patients before surgery, but was reduced to 20% and 5%, respectively, 6 months after surgery. The differences between the two groups in the scores for the three indexes did not did not reach statistical significance before and immediately after the operation, but the differences in the scores after 6 months and immediately after surgery in the control and surgical groups were significant at levels of 2.5% for the withdrawal index, 5.0% for the developmental index, and 1% for the attention deficit disorder index (Fig. 2).

Self-mutilation behavior (mainly head-banging) improved a few months after surgery in 9 of the 10 patients who had exhibited this symptom. One patient switched from banging the head to banging the face. The panic and irritability noted in 35 of the 65 patients disappeared or markedly decreased after surgery (Table 1). The periods during which panic or irritability were displayed became shorter in duration, and the patients were able to tolerate and obey commands not to engage in certain activities.

Seven of the 8 patients with motor dysfunction improved after surgery (Table 1). They became better able to engage in age-appropriate sitting, walking, jumping, and tricycle-riding within a few months after the operation. Only 1 patient continued to exhibit the same degree of physical awkwardness after the surgical procedure.

Among the 49 patients in whom D.Q. was chronologically evaluated pre- and postoperatively, an upward trend was seen in 20 (41%), especially among those who had a D.Q. score >60 preoperatively, and a downward trend in 29 (59%) (Fig. 3). Thus, in terms of overall results, 42 of the 65 patients improved markedly, and 15 were able to enter regular kindergarten or elementary school classes. Nineteen improved slightly after surgery, while 4 did not exhibit any change.

CT and MRI findings

The results of CT and MRI examinations conducted 2 months after surgery showed that the frontal lobes had increased in size in all 65 patients (Fig. 7). Three-dimensional CT images showed an increase in volume of the frontal calvarium and anterior fossa (Fig. 4). The area of bone trimmed during the surgical procedure was gradually reconstructed with neobone over time. Older patients (more than 6 years of age) tended to form neobone more slowly than younger ones. One year after surgery only 4 of the 57 patients in this group had produced sufficient neobone to compensate for 80% of the postsurgical bone defect, but by 2 years after surgery 10 of 12 patients had done so. In 2 patients (both 7-year-old boys) who had produced some neobone, it compensated for less than 50% of the bone defect.

SPECT findings

Postoperative SPECT examination was performed in 54 of the 65 patients. Among the 41 who were found to have reduced CBF before surgery, 39 exhibited improved flow 2 months after surgery, and 2 exhibited no change. In the 13 who had normal CBF prior to surgery, it remained normal afterward.

Surgical complications

Small contusions were evident after surgery in 2 patients, and massive bleeding from the superior sagittal sinus occurred in 1 patient. One boy on whom surgery was performed at 8 months of age underwent reoperation at the age of 2 years (double white circle in Fig. 3) because of speech problems and frontal lobe constriction that was detected in neuroradiological examinations. There were no perioperative infections in this series.

Discussion

Trigonocephaly is divided into two types: nonsyndromic with no brain anomalies and syndromic in which anomalies are present. All of our patients had the nonsyndromic type with no brain abnormalities. All had mild trigonocephaly. Although it has been thought that in mild cases there are no manifest clinical symptoms, we have long been concerned about the relationship between clinical symptoms and mild trigonocephaly. Although Anderson et al. [2], Shilito and Matson [25], and Oi and Matsumoto [19, 20] suggested that a high proportion of patients with typical trigonocephaly might be mentally retarded, it was apparent at the time of a report by Collmann et al. in 1996 [6] that a consensus had been reached in the field of pediatric neurosurgery on trigonocephaly and clinical symptoms.

On the other hand, Sidoti et al. [28] reported in 1996 that cognitive and/or behavioral abnormalities were present in a significant number of patients with metopic synostosis and that the incidence of such abnormalities appeared to be higher in patients over 6 years of age. The 1998 report of Kapp-Simon [15] also described a higher rate of retardation among children with metopic synostosis, at a similar level to that reported by Sidoti et al. [28]. Bottero et al. [5] found that 31.6% of their surgical patients had evidence of some degree of developmental delay. These reports were concerned with patients who had typical trigonocephaly with the keel-shaped deformity of the forehead. Bottero et al. [5] reported that 23% of nonsurgical patients with the mild form of trigonocephaly manifested developmental delay. All of our patients had the mild form with heel-shaped deformity of the forehead, despite the fact that 7 could be classified as having the severe form according to Oi and Matsumoto's grading system [19, 20]. In agreement with Collmann et al. [6], we have noted that many children with normal mental development have a metopic ridge (mild trigonocephaly), and surgery is not indicated for these patients. However, we have also treated patients with mild trigonocephaly associated with clinical symptoms such as delays in language development, hyperactivity, impaired social interaction, motor dysfunction, etc. who improved after surgery.

Most of our patients developed normally until they were 1 year old, and then they clearly began to manifest developmental delays with increasing age. Some even regressed in terms of language acquisition and use. These symptoms can make the diagnosis of mild trigonocephaly difficult. The most characteristic facial feature of mild trigonocephaly in our series of 65 patients was the heel-shaped deformity of the forehead. The temples were also depressed, as seen in typical cases, but to a lesser degree. Hypotelorism is difficult to identify on the basis of physical appearance, but is clearly revealed on skull X-ray and 3D-CT images. The most important physical sign was the palpable, but not abnormally high, metopic ridge in all of our patients. After our initial experience of treating patients with mild trigonocephaly was explained to the pediatricians working at an institution for children with developmental delays, they were able to diagnose the condition by palpation of the fore-

head and subsequently referred numerous patients to us. Our institution is a general hospital, and therefore we perform surgery in only one or two cases of craniosynostosis each year. Thus, it is important for neurosurgeons to maintain contact with pediatricians or pediatric psychiatrists who treat patients with developmental delays.

The final diagnosis should be made from 3D-CT images, which clearly show a metopic ridge, depressed pterional regions, hypotelorism, and small anterior fossae. We have also been using 3D-CT to diagnose other forms of craniosynostosis, because this imaging modality clearly shows the fusion of sutures.

Even though our patients had the mild form of trigonocephaly, frontal stenosis was apparent on calculation of the IPD/ICD [5] ratio on standard axial CT images. Standard axial CT and MRI revealed small frontal lobes, with no abnormalities in the brain. MRI should be performed to rule out an association with any type of brain anomaly, which is often seen in this disease, especially the syndromic type [6]. Preoperative SPECT in our patients showed decreased CBF, mainly in the frontal lobes, in approximately 76%. SPECT has been used to evaluate CBF in patients with various types of craniosynostosis, and decreased regional CBF was found in 62–86% of patients [7, 24, 29]. The neuroradiological findings in our patients appeared to contribute to the dysfunction of the frontal lobes.

Our surgical procedure is a combination of that of Marchac [16] and Raimondi and Gutierrez [21]. The removal of the supraorbital bar included bilateral “roofs” to open the frontosphenoidal and frontoethmoidal sutures. Raimondi and Gutierrez [21] stated that the lesser and greater wings of the sphenoid bone must be removed to correct the coronal synostosis. This procedure allows the frontal and temporal lobes to expand. We used the above two methods to treat mild trigonocephaly, because during surgery the brain always appeared tight and pulsating with pressure after the cranial vault was opened. Another reason for performing this procedure is the hypothesis that the cause of craniofacial dysmorphisms is synostosis occurring in the sutures of the cranial base, particularly in the frontosphenoidal and frontoethmoidal sutures in the anterior cranial fossa [14, 16, 17, 23]. After surgery, the patients’ foreheads were flat, the temples were no longer depressed, and, interestingly, the space between the eyes was wider.

The clinical symptoms in our patients improved to some degree after surgery compared with those in the 15 nonsurgical controls. One of the most impressive changes after surgery was in the comprehension of language, which might also contribute to an improvement in their language acquisition and use. Of the 61 patients who spoke few or no words before surgery, 37 increased their vocabulary after surgery, but in patients who could speak more than 10 words before surgery there was a vocabulary increase of a mean of 84% after surgery.

The JCBCL was used to analyze changes in pre- and postoperative behavior, and the results were compared with those in the nonsurgical group. The withdrawal (mainly autistic tendencies) and attention deficit disorder (mainly hyperactivity) indexes improved markedly soon after surgery. It is important for these patients to undergo rehabilitation therapy to allow them to study and learn in the school setting in the future.

The self-mutilation, panic, and irritability ceased or decreased markedly in most patients after surgery. The cessation of head-banging was especially appreciated by family members. Motor dysfunction also markedly improved after surgery. It is therefore thought that symptoms such as delay in language development, motor dysfunction, hyperactivity, autistic tendencies, and self-mutilation are related to the frontal lobe dysfunction [3, 4, 13, 18].

Regarding the D.Q., our previous investigation [26] found an increasing trend among surgically treated patients that was greater than among a general group of patients with mental retardation. In the present patient series, an upward trend was also noted, although those with a lower D.Q. before surgery did not improve as much afterwards as those with a higher presurgical D.Q. Previous investigations [5, 15, 28] in patients with trigonocephaly attempted to determine whether early surgery could prevent the development of mental delay. Our patients already exhibited mental delay preoperatively, so that we cannot compare our results with those in previous studies. However, we think that the placement of 15 of the 65 patients in regular school classes should not be ignored.

Postoperative neuroradiological findings included expanded frontal lobes as shown on standard axial CT and MRI. The depressed temples seen on 3D-CT also became flatter. Postoperative SPECT demonstrated improvement in CBF in the frontal lobes in about 95% of patients. Other researchers have reported that postoperative SPECT in craniosynostosis patients also showed increased CBF in 70–100% [7, 24, 29]. David et al. [7] noted that asymmetric CBF before surgery in 5 of 7 patients with single-suture craniosynostosis became symmetric after surgery. They saw this improvement in CBF as supporting early surgical intervention to prevent potential compromise of the central nervous system secondary to abnormal CBF.

In our study, the MRI findings showed that all patients had mild trigonocephaly and none had cerebral malformations. The indications for surgery in mild trigonocephaly were earlier limited to prophylactic/cosmetic extracranial frontal ridge resection, as advocated by Bottero et al. [5] and Collmann et al. [6]. Currently, it is still generally thought that the surgical indications for mild trigonocephaly are extremely limited. In addition, it is recommended that patients with typical trigonocephaly undergo surgery when less than 1 year of age, since better results can be expected than in children who are older than 1 year at the time of surgery [22]. All of our 65 pa-

tients exhibited clinical symptoms despite the diagnosis of mild trigonocephaly, and most underwent surgery after 1 year of age because the condition is difficult to diagnose prior to that. Based on our surgical results (approximately 94% overall improvement), it is recommended that surgery be performed in patients with mild trigonocephaly who exhibit clinical symptoms even when they are older than 1 year of age.

The improvement of clinical symptoms due to the release of constricted frontal lobes in our patients with mild trigonocephaly was documented by pre- and postoperative neuroradiological findings. The expanded frontal lobes after surgery allowed the patients to learn and increase their understanding of their surroundings, making them easier to teach. However, prognostic factors that might indicate which patients will have good surgical outcomes or to what degree symptoms can be expected to improve are still unknown. Further studies are therefore required.

In conclusion, it should be emphasized that the most important diagnostic sign of trigonocephaly is the metopic ridge when the forehead is palpated. We uggest that a high proportion of children with developmental delay have mild trigonocephaly, and that in most patients the delays and other symptoms can be reduced by decompressive surgery.

Acknowledgement We would like to express our thanks to Okinawa Prefectural Officer Kouzou Tamori for analyses of our statistical data.

References

1. Achenbach TM (1992) Manual for the child behavior checklist 2–3 and 1992 profile. Department of Psychiatry, University of Vermont, Burlington, Vt
2. Anderson FM, Gwin JL, Todt JL (1962) Trigonocephaly: identity and surgical treatment. J Neurosurg 19:723–730
3. Barkley RA, Grodzinsky G, DuPaul GJ (1992) Frontal lobe functions in attention deficit disorder with and without hyperactivity: a review and research report. J Abnorm Child Psychol 20:163–188
4. Baron-Cohen S, Ring HA, Wheelwright S, et al (1999) Social intelligence in the normal and autistic brain: an fMRI study. Eur J Neurosci 11:1891–1898
5. Bottero L, Lajeunie E, Arnard E, Marchac D, Renier D (1988) Functional outcome after surgery for trigonocephaly. Plast Reconstr Surg 102:952–958
6. Collmann H, Sorenson N, Krauss J (1996) Consensus on trigonocephaly. Child's Nerv Syst 12:664–668
7. David LR, Wilson JA, Watson NE, Argenta LC (1996) Cerebral perfusion defects secondary to simple craniosynostosis. J Craniofac Surg 7:177–185
8. Delashaw JB, Persing JA, Park TS, Jane JA (1986) Surgical approaches for the correction of metopic synostosis. Neurosurgery 19:228–234
9. Dhellemmes P, Pellerin P, Lejeune JP, Lepoutre F (1986) Surgical treatment of trigonocephaly. Experience with 30 cases. Child's Nerv Syst 2:228–232
10. Di Rocco C, Velardi F, Ferrario A, Marchese E (1996) Metopic synostosis: in favour of a "simplified" surgical treatment. Child's Nerv Syst 12:654–663
11. Dominguez R, Oh KS, Bender T, Girdany BR (1981) Uncomplicated trigonocephaly. A radiographic affirmation of conservative therapy. Radiology 140:681–688
12. Friede H, Alberius P, Lilja J, Lauritzen C (1990) Trigonocephaly: clinical and cephalometric assessment of craniofacial morphology in operated and nontreated patients. Cleft Palate J 27:362–367
13. Gedye A (1991) Extreme self-injury attributed to frontal lobe seizures. Am J Ment Retard 96:81–85
14. Hoffman HJ, Hendrick EB (1979) Early neurosurgical repair in craniofacial dysmorphism. J Neurosurg 51:796–803
15. Kapp-Simon KA (1998) Mental development and learning disorders in children with single suture craniosynostosis. Cleft Palate Craniofac J 35:197–203
16. Marchac D (1978) Radical forehead remodeling for craniostenosis. Plast Reconstr Surg 61:823–835
17. Moss ML (1975) Functional anatomy of cranial synostosis. Child's Brain 1:22–23
18. Ohnishi T, Matsuda H, Hashimoto T, Kunihiro T, Nishikawa M, Uema T, Sasaki M (2000) Abnormal regional cerebral blood flow in childhood autism. Brain 123:1838–1844
19. Oi S, Matsumoto S (1986) Trigonocephaly (metopic synostosis). Clinical, surgical and anatomical concepts. Child's Nerv Syst 3:259–265
20. Oi S, Matsumoto S (1986) Early radical operation for trigonocephaly in infancy – pathophysiological concepts for and operative procedure for closure of metopic suture. No Shinkei Geka 14:1087–1092
21. Raimondi AJ, Gutierrez FA (1977) A new surgical approach to the treatment of coronal synostosis. J Neurosurg 46:210–214
22. Renier D (2000) Craniosynostosis: surgical techniques according to age and late results. Shouni No Nousinkei 25:1–11
23. Seeger JF, Gabrielsen TO (1971) Premature closure of the frontosphenoidal suture in synostosis of coronal suture. Radiology 101:631–635
24. Sen A, Dougal P, Padhy AK, Bhattacharya A, Kuma R, Bal C, Bajpai M, Mitra DK, Basu AK (1995) Technetium-99m-HMPAO SPECT cerebral blood flow study in children with craniosynostosis. J Nucl Med 36:394–398
25. Shilito J Jr, Matson DD (1968) Craniosynostosis: a review of 519 surgical patients. Pediatrics 41:829–853
26. Shimabukuro S, Shimoji T, Sugama S (2001) Cranioplasty to isolated trigonocephaly with developmental disturbance. No To Hattatsu 33:487–493
27. Shimoji T, Yamada M, Hara S (2000) Trigonocephaly associated with symptoms – mainly nonsyndromic type (in Japanese). Shouni No Nousinkei 25:43–48
28. Sidoti EJ Jr, Marsh JF, Marty-Grames L, Noetzel MJ (1996) Long-term studies of metopic synostosis: frequency of cognitive impairment and behavioral disturbances. Plast Reconstr Surg 97:276–281
29. Uemura T, Noda K, Hayashi T, Sato K (1998) Evaluation of craniosynostosis surgery: Technetium-99-m-HMPAO SPECT cerebral blood flow study in children with craniosynostosis. J Jpn Plast Reconstr Surg 18:489–496