Mild trigonocephaly
-Report of 300 operative cases-
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Abstract Introduction: Since 1999, we have reported mild trigonocephaly with symptoms mainly accompanied with developmental delays. We would like to report the operative results of 300 patients.
Patients and methods: All patients had some kind of clinical symptoms. The diagnosis made recognizing ridge of the fused metopic suture by palpation and a three-dimensional computed tomography (3D-CT). Most patients (287 among 300) could not be diagnosed during infants because of the mild changes of their facial features and their natural development as infants. They suffered from symptoms such as mental retardation, delayed language development, hyperactivity, autistic tendencies, panic and irritability, motor dysfunctions, self-mutilation and sleeping disturbances. Two hundred thirteen patients were recorded intracranial pressure (ICP). Developmental quotients (D.Q) were also measured.

Results: Two hundred fifteen (73.6%) of 292 patients with delays of language development were improved within a year after surgery. Other symptoms also improved: 91.6% in hyperactivity, 76.5% in autistic tendencies, 92% in panic, 90% in motor dysfunctions 88.6%, in self-mutilation and 92% in sleeping disturbances. The chronological scores in D.Q maintained parallel in most patients. Measurements of ICP resulted in 10 cases (5%) under mean 10 mmHg, 33 cases (15%) between 11 to 15 mmHg and 170 cases (80%) over 16 mmHg. The pulse pressure calculated mean 9.5mmHg. Decompressive cranioplasty was applied to all patients.
Conclusion;
i) Patients with mild trigonocephaly may have a possibility to show clinical symptoms.
ii) Since the ICP seems to be high, decompressive cranioplasty may be a reasonable treatment for these patients.

Key words
Mild trigonocephaly,
Developmental delay,
Developmental quotient,
Three-dimensional computed tomography,
Intracranial pressure,
Decompressive cranioplasty

Introduction

In the patients with typical nonsyndromic trigonocephaly, mental deficiency cannot serve as an argument for brain constriction, since it is a common feature in syndromic trigonocephaly, probably because of coincidental cerebral malfunction (8). From this point of view, it is generally believed that an indication for surgery to typical trigonocephaly has been applied for cosmetic and long term psychological effects (2) (8) (14) (15). However, in

recent years, some papers have been published that patients with the single-suture synostosis might present neuropsychological dysfunctions including global intelligence (11) 12) (13) (25) (26). They also stated that the risk of poor neurobehavioral outcome in children with single-suture synostosis may be associated three to five times higher than average (13) (26). There were other reports of even higher incidences of neurobehavioral abnormality (3) (25). These reports were focused on typical single-suture craniosynostosis. It seems that mild types of craniosynostosis such as we had reported before (22), were not involved. Generally, in terms of mild nonsyndromic trigonocephaly, surgical indication was not considered (8) (15). However, the results of ours which we had published before (22) (23), were quite different from these general ideas. Although we cannot explain the relation between the symptoms and mild deformities of brain as Speltz et al (26) reviewed, we have found many cases with clinical symptoms in the mild trigonocephaly. In most of these cases, high intracranial pressure was seen. So that we have reported decompressive cranioplasty is necessary as a treatment (22) (23). Finally we have 300 consecutive patients who underwent surgery. The purpose of this paper is to report the clinical features and treatment with mild nonsyndromic trigonocephaly.

Patients and methods

Patients

From October 1994 to June 2006, 300 children diagnosed with mild trigonocephaly. They were mainly examined and treated at Okinawa Prefectural Naha Hospital and our hospital. The age distribution was 2-8 (mean 5) years old. The sex ratio was boy dominant, 230 boys and 70 girls (Fig. 1). All 300 patients were considered to be included in the category of mild nonsyndromic trigonocephaly. All of them underwent chromosomal testing with no abnormal findings. Although they were categorized as

“nonsyndromic”, they had suffered with their clinical symptoms. We have explicitly informed the parents that this surgery was for the purpose of improving some of the child’s symptoms, rather for cosmetic purposes, since several cases were experienced with remarkable improvement of their clinical symptoms. We operated after accepting getting parental consent for surgery.

Preoperative evaluation

Facial features

Our patient’s foreheads typically resembled a heel more than the usual keel shape seen in the typical trigonocephaly. The metopic ridge was easily palpable and visible in some in the moderate cases. The temples were depressed bilaterally. Hypotelorism was not identified physically, but it could be noted on plain X-Ray and 3D-CT. Milder cases were more difficult to diagnose on the basis of physical examination due to the shallower metopic ridge flatter forehead in those patients. The diagnosis was therefore based on 3D-CT.

Symptoms

Only 13 out of 300 patients could be noted before the age of 1 year by their symptoms and their features. Seventy two patients had regressions in language acquisition and use or their development, which mainly occurred during their one year of age. All patients had some type of clinical symptoms as follows. Main symptoms were global delay in language development, hyperactivity, autistic tendency, self-mutilation, panic and/or irritability, motor dysfunction and sleeping problems (Table 1). In 292 of the patients, language development was delayed at different stages; in forms ranging from an inability to utter meaningful words to being unable sustain a conversation (Table 2). Hyperactivity and/or attention deficit/hyperactivity disorder

was noted in 225 of the patients. The related symptoms included excessive movement, climbing behavior, wandering away from parents and difficulty in sitting still. One hundred eighty seven patients were noted to have autistic tendency, manifested by difficulty in eye contact, sticking to special things, stereotyped and repetitive motor mannerisms and aloofness and lack of social or emotional reciprocity. One hundred seventeen patients exhibited panic and irritability when they are stopped from doing what they wanted. They kept screaming and crying for a long time. Seventy three patients had motor dysfunction, manifested as not being able to sit, jump, or ride a tricycle at the appropriate ages and physical awkwardness. Seventy patients noted to have self-mutilation, mostly in the form of head banging. After January 2005, we had added a question while taking history whether the child had sleeping disturbance or not. Twenty nine patients out of 90 (32.2%) suffered from it. Some of them woke up during the middle of the night and kept crying loudly for a long time. Some found to be quite difficult to go to sleep. Most patients were thought to be mentally retarded. Their DQ was evaluated by the K-form Developmental Test (a commonly used test in Japan) (Table 3). Twenty patients were over 80 points in DQ. They all had social problems except one who presented only severe headaches.

Neuroradiological findings

The skull X-rays were taken in all patients. The digital markings were noted over 3/4 areas in the lateral views in 214 patients. (The definition of the areas was reported before (23).) The finding of the metopic ridge shown by the 3D-CT scans was considered to indicate the final diagnosis for this mild trigonocephaly. 3D-CT was used to classify the severity of this disease. In our cases, 90 cases were considered as mild and 210 cases were considered as moderate by Oi’s classification (19). Regular CT and magnetic resonance imaging (MRI) applied to all patients to evaluate brain

parenchyma. No abnormal findings were morphologically seen in all patients. Two hundred forty six patients underwent preoperative single-photon emission tomography (SPECT) with technetium-99m ethyl cysteinate dimer and 160 patients were pointed to have decreased cerebral blood flow (CBF), mainly in the frontal lobes. We must note that these must be regarded as qualified results, since there is no well accepted standard measurement using SPECT for the evaluation of CBF in childhood.

Measurement of ICP

Intracranial pressure was tried to measure in the consecutive patients after 50^{th} case. We succeeded in 213 cases to obtain feasible data. All of the patients underwent the protocol described below. The patients were placed under endotracheal general anesthesia using Sevoflurane. The maximal inspiratory pressure was controlled at around 18 mmHg. ICP was measured after a burr hole had been made and a sensor (Camino multiparameter monitoring system, Integra NeuroSciences, U.S.A.) was inserted in the epidural space on the frontal lobe just anteriorly of the right coronal suture. The first recordings were made at a PCO_2 level of around 30 mmHg for neuroanesthesia (hypocapnea), and the second at a PCO_2 of around 40 mmHg as in natural breathing (normocapea). The blood gasses were examined at each pressure condition.

Surgical procedure

The procedure might be referred in detail to our previous report (22). It should be emphasized that the important points are sufficient decompression of the major and lesser sphenoid bone including sphenoid ridge bilaterally and the supraorbital bar including roofs removed in one piece to open the frontosphenoidal and frontoethomoid sutures.

During the surgery we had an impression that the very thin and wide sphenoid ridge was sharply sticking between the frontal and temporal lobes towards the dura of the sylvian fissure.
The procedure should be referred to as decompressive craniotomy. Since some patients had a remaining large bone defect, we made a minor change of the procedure. After case number 95 which underwent surgery at January, 2001, a piece of bone was placed on the central frontal area (Fig 2).

Results

Facial features

After surgery, the heel-like appearance of the forehead was improved. It became broad and flat in almost all of the cases. Also the depressed temporal area seemed to be released.

Symptom

Post-operatively, symptoms were evaluated between six months and one year after surgery.
All of the factors seemed to be improved. Two hundred fifteen out of 292 (73.6%) in language delays, 206 out of 225 (91.6%) in hyperactivity, 143 out of 187 (76.5%) in autistic tendencies, 108 out of 117 (92%) in panic and irritability, 66 out of 73 (90.4%) in motor function disorder, 62 out of 70 (88.6%) in self-mutilation and 27 out of 29 (93%) in sleeping disturbance respectively (Table 1). Language delays were precisely investigated one year after surgery (Table 2). Forty four out of 78 (56%) patients who could not express meaningful words before surgery remained in the same condition after surgery, but others (44%) had progress to express some words, sentences and even conversation. Patients who used fewer meaningful words prior to surgery had a tendency to show less improvement.
One hundred ninetyone patients were evaluated pre- and

post-operatively by a K-test (Table 3). One hundred eighteen patients (62%) remained at their post-operative DQ scores between minus to plus 10 points. Six patients (3%) dropped their DQ below 20 points; however 23 patients (12%) showed a remarking increase more than 20 points. There is no statistical significance between them.

One hundred thirty six patients among these could be evaluated chronologically more than one year (Fig. 3). It was clearly seen in this follow up data that most of patient's scores were almost running horizontally, although some worsening and improvement were seen. Additionally, a total of 44 patients were able to enter regular kindergarten or elementary school classes.

Here we would like to present two example cases to show what kind of patients we are treating.

Case 1

This four year old boy grew up normally until his one year of age, there was no developmental delays in speech and no abnormality in behavior. After 1.5 years of age he regressed in language and behavior. At the time he came to our hospital, he was not able to speak meaningful words, not able to point his finger to what he wants, kept walking around, and not playing with other children (Fig. 4 a). His 3D-CT showed moderate trigonocephaly and the skull X-Ray showed marked digital markings (Fig. 4 b). He underwent surgery in Oct. 2005. ICP measurement evidenced mean 19 mmHg under PCO_2 41.5 mmHg (Fig. 4 c). Four months later he could spontaneously speak words, learn words using cards with his father, and play with his mother (Fig. 4 d). At present, two years post-operation, he can speak sentences and play with other children at kindergarten. However, he was not able to enter regular elementary school due to his intelligence level. We estimated improvement in his language, hyperactivity and autistic tendency.

Case 2

A six year old girl with severe mental retardation was referred to us and diagnosed as a "mild" trigonocephaly. She had been

loosing her acquired words. She could obey to only simple orders. She had been moving all the time without any eye contact. 3D-CT showed a metopic ridge and the “mild” degree of trigonocephaly. The skull X-Ray showed no digital markings at all (Fig. 5 a). Preoperative SPECT reported no finding of decreased CBF. She had surgery at six years of age in Jan 2004. During surgery, ICP was recorded and had the mean ICP of 18 mmHg under PCO_2 42.3 mmHg (Fig. 5 b). At one year post-operation, she spoke two-word sentences, was no longer losing newly acquired words, and her hyperactivity was notably subsided. She could understand her mother’s words much better than pre-operation. She had been getting along better with other children in her special school. We estimated her that she had slightly improved because of her improved language, understanding and behavior.

Measurement of ICP

The mean PCO_2 was 31.1 mmHg during hypocapnea and 39.7 mmHg during normocapnea. The values are close to those designed. During normocapnia, the ICPs were recorded less than 10 mmHg in 10 patients (5%), 11 to 15 mmHg in 33 patients (15%) and more than 16 mmHg in 170 patients (80%). The pH recorded 7.44 during hypocapnea and 7.36 during normocapnia. ICP during normocapnea became higher in all patients (Table 4). The mean ICP and pulse pressure in each group showed 8.9 and 8.9 mmHg in the first group, 13.6 and 8.6 mmHg in the second, and 21.5 and 9.8 mmHg in third respectively (Table 5).

Neuroradiological evaluation

Post-operative 3D-CT and MRI showed larger frontal lobes and anterior fossae (Fig.2) in all patients. Post-operative SPECT examination was performed one year after surgery, but it could not be evaluated precisely because bone defects still existed in the central frontal area.

Surgical complication

As surgical complication, there are two small contusion in the frontal lobes in 2 patients, one massive bleeding from superior sagittal sinus during surgery and one intracerebral hematoma due to a trauma in a week after surgery which needed a surgical intervention. Two patients had a cranioplasty after 5 and 6 years from surgery because a large bone defect remained in the forehead. One boy whom surgery was performed at 8 months of age underwent a second operation at the age of 2 years because of frontal lobe constriction and developing sagittal synostosis.
Cell saver (ELECTA, Dideco S.r.l., Italy) has been used since April 2004 (from case number 171). Only two (1.5%) patients had a blood transfusion since then.
There were no surgical infections in this series.

Relationship between clinical results and physical examinations

It was difficult to put patients making a score how much they were improved due to their various symptoms. Thus, the patients' clinical results were subjectively divided into unchanged (estimated 18 patients), slightly improved (130 patients) and improved (152 patients). Pre-operatively, the unchanged group of patients tended to have a lower DQ, lesser findings of digital markings on skull X-ray (lately identifying by 3D-CT), no abnormal findings on SPECT and mild degree of trigonocephaly. In contrast, the improved group of patients tended to be younger, to have relatively higher DQ, marked digital markings, and decreased CBF on SPECT and moderate degree of trigonocephaly. Regarding ICP, the mean value of the ICP was 23.7 mmHg in the unchanged group, 19.6 mmHg in the slightly improved group and 19.2 mmHg in the improved group respectively. In the unchanged group, 11 out of 18 patients underwent this monitoring. In these 11 patients, the ICP was less than 10 mmHg in one case. Three

patients were in between 15 to 18 mmHg 7 cases were more than 25 mmHg (Table 6). However, none of these indicators statistically influenced to the clinical results.

Discussion

It is generally accepted that patients with nonsyndromic single-suture craniosynostosis manifest mental retardation in a low rate (2.4 to 4.8%) (21). In the field of pediatric neurosurgery, as statement of Collmann (8) in 1996, there appears to be a consensus that there are no manifest clinical symptoms in cases of typical trigonocephaly and a surgical indication is to be cosmetic (14) (15). However, several reports (5) (11) (25) regarding symptoms in typical trigonocephaly since 1996 presented that patients had mental retardation and developmental delays in higher incidences than it was thought. All of our cases were considered to be nonsyndromic and the severity of trigonocephaly was mild. We ourselves also did not believe that a mild trigonocephaly will come out with clinical symptoms at all until 1994. Our first case, a two year old boy with hyperactivity and no meaningful words who was classified as moderate trigonocephaly, had surgery for cosmetic reasons in 1994. Shortly after discharge, His mother reported that he was speaking and being subsided with his hyperactivity. Since then we have had almost the same experience in next several cases. The results were the same, all children were improving. All patients were mild cases, and their ages were over one year old except one case. After these experiences, we reached to an idea that patients with mild trigonocephaly may have clinical symptoms and may need treatment for their symptoms, not for cosmetic purposes. In 1999 we began to present our cases at academic conferences and also we published a report to this journal in 2002 (22) and 2004 (23).
Regarding a type of mild craniosynostosis, difficult to diagnose by their head shape like ours, several reports presented clinical symptoms in older patients and discussed about the elevated ICP

(7) (17) (27). Their cases presented with symptoms of raised ICP, mainly papilledema. In their cases, a single-suture craniosynostosis was involved. Martinez-Lage et al. (17) referred to these as minor forms of “occult” craniosynostosis. Our cases also presented their symptoms at older than one year of age, except two cases. Most cases started to show their symptoms after one and a half years of age. 25% of the patients had regression in language acquisition and use or their development, which mainly occurred during their one year of age. At this time we have noticed that patients with mild trigonocephaly were developing normal during infancy. When we diagnose an infantile patient with mild trigonocephaly, we inform their parents that close observation is needed because of the reasons above. Bottero et al. (5) already pointed out in 1998 that children with a minor form of trigonocephaly not warranting surgical intervention had developmental delays in 23% of their database. Regarding symptoms, the previous reports (3) (5) (11) (25) pointed to the presence of symptoms in patients with typical trigonocephaly described merely mental retardation, cognitive and/or behavioral abnormalities. Our first paper (22) described cases of surgically treated mild trigonocephaly and their clinical symptoms precisely. Compared with fifteen non-surgical cases, our surgical cases had superior results statistically. Again, in this report of three hundred cases, most of them presented with mental retardation associated with language delay, hyperactivity, autistic tendency, panic and irritability, motor function disorder and self-mutilation. It was already described in the previous paper how much these symptoms were improved. Every symptom got better in high incidences compared to pre-operative state, even though the number of patients was increased to three hundred. This time sleeping disturbance was added. We could estimate about 25% of patients had difficulty in sleep such as follows. Some never fall into sleep until late at night, some woke up in the middle of the night crying loudly, and some got up early in the morning and remained unsettled for a long time. These disappeared soon after

surgery in 92% of patients.

It is difficult to evaluate mental retardation and/or cognition in a child. However, a score of DQ obtained by K-form test will be, not absolute, but may be one of the indicators. The results of DQ score have not change in 62% of patients. These were more apparent in chronologically followed tests in 136 patients after surgery (Fig 3). Several patients were dramatically showing progress in their scores. In general, scores of most mentally retarded patients has a declined score as they grow up. Considering this fact it may be better to emphasize that our patients after surgery were keeping their DQ scores unchanged.

The digital markings on skull X-Rays were noted in many patients (more than three-fourths on the lateral skull X-Rays in 70% of patients). There were a couple of reports to show the relationship between high ICP and the digital markings in craniosynostosis (1) (29). Agrawal et al. (1) reviewed their postoperative patients with sagittal synostosis and stated that patients with marked digital markings on the skull X-Rays tended to have high intracranial pressure and symptoms such as headaches, head banging and irritability.

In our early cases without ICP monitoring, when we touched the brain itself over the dura matter there was an impression that it was tighter than normal cases. The high incidences of digital markings and impression of high ICP during surgery made us start recording ICP after consecutive number of 50^{th} patient. ICP was recorded in the same manner in all patients as described. In this way, the recorded ICP values were thought to be close to actual regional values. ICP recording, PCO_2 was controlled around 40 mmHg, resulted below 10mmHg in 10 (5%), between 11 to 15 mmHg in 33 (15%) and over 16 mm Hg in 170 (80%) patients. The ICP and pulse pressure increased parallel with the increase of PCO_2. These results might be concluded high ICP and low intracranial compliance in most of our cases. As our latest report (23) postulated and referring the report of Inagaki et al. (10) treating almost the same kind of patients, it could be said that

most mild trigonocephaly patients with clinical symptoms had suffered from raised ICP, although we also accept that the method way of epidural recording of ICP only for several minutes was not accurate compared to the continuous recording. In the past, there were many reports regarding measurement of ICP in patients with craniosynostosis which stressed the importance of deciding indication for surgery (6) (20) (28). So that, it may be appropriate to say that the application of decompressive cranioplasty in our cases may be reasonable from a point of view of increased ICP. It is important to manipulate the anterior part of skull base which results to produce larger anterior fossae (18). It also contributes to make the frontal lobes increasing in size and even cosmetically better feature.

Analyzing our clinical indicators, there were tendencies in improved groups to be younger, to have relatively higher DQ, marked digital markings, moderate degree of trigonocephaly and decreased CBF on SPECT. However, as shown in the second case report, some patients with lower DQ, milder degree of trigonocephaly, lesser findings of digital markings and normal CBF on SPECT improved postoperatively. All patients in the unchanged group were estimated as severely retarded in pre-operation; however, four patients who tried K-form test with resulting not completed, namely point 0, made remarkable improvement after surgery. It is interesting that most patients in the unchanged group had high ICP. From all these results, it is thought that there are no absolute clinical indicators for indication for surgery.

As several papers pointed out, patients with nonsyndromic single-suture craniosynostosis might develop mental retardation and learning disorders (5) (11) (25). After these reports, Kapp-Simon et al. (12) (13) and Speltz et al. (26) extensively reviewed neurodevelopment of children with single-suture craniosynostosis. In the same period, several reports published regarding mental development, speech, cognitive disorders and behavioral problems in patients with single suture

craniosynostosis (3) (4) (9) (16) (24) (30). These reports were all for typical craniosynostosis not to be mild one which did not require for cosmetic procedure. Kelleher et al. (14) (15) described that patients with trigonocephaly presented with attention deficit disorder, autism and hyperactivity. They included patients with mild degree trigonocephaly, but they postulated that there was no indication for surgery on them. We operated on patients with mild trigonocephaly associated with clinical symptoms and postulated some surgical befits on them (22) (23). This subject had been presented in academic conferences in Japan and international meetings since 1999, but it was very difficult to be accepted. The patients' facial features are as minor as well as radiological findings. It is natural to doubt this postulation. However, in Japan a few institutions started to operate on patients with clinical symptoms associated mild trigonocephaly. Inagaki et al. (10), from one of these, reported 36 patients with clinical symptoms such as ours and mild form of craniosynostosis. They operated on 25 patients after confirming high ICP. Also all of their cases had shown improvement in certain degree and in some dramatically.

We have now analyzed 300 cases; however we could not find any specific indicator to decide surgical indication besides the patients' clinical symptoms and the metopic ridge on 3D-CT.

In spite of our institution is located on such a small island, many patients (204 cases) with mild trigonocephaly were found among mentally retarded group and mild development delays (speech delay, hyperactivity and autistic tendency and so on). Other patients included three American-Japanese, one Chinese-Japanese, and ninety three patients from main islands in Japan. We suppose that this kind of patient exists around the world and is hopefully investigated as soon as possible.

In conclusion, a certain number of patients with nonsyndromic type of mild trigonocephaly apparently manifest clinical symptoms in our experience. Although it is not clear that all of the symptoms are directly connected with mild trigonocephaly, it is apparent in our experience that this condition is at least making

the patients' symptoms worse. As we found that the patients with mild trigonocephaly recorded high ICP in the most cases, the appropriate treatment might be decompressive cranioplasty.

Finally, we would like to postulate to all the physicians who are related to child healthcare. When one sees a child with development delays such as language delays, hyperactivity and autistic tendency, consider this child may have this symptom due to increased ICP related to mild trigonocephaly. It is simple; just touch the child's forehead. If there is a palpable ridge, there might have a possibility to have an improvement at least in some degree by undergoing decompressive cranioplasty.

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Legends

Table 1; improvement in symptoms of mild trigonocephaly after surgery (n=300)

Table 2; changes in language use and acquisition from before to after surgery (n=292)

Table 3; changed DQ score after surgery

Table 4; blood gas analysis and intracranial pressure (ICP) levels
The results are the mean of 213 patients during hypocapnea and normocapnea

Table 5; ICP and blood gas levels during normocapnea

Table 6; post-operative outcome and physical examination results
UC; unchanged, SI; slightly improved, I; improved

Fig. 1; age distribution and sex of patients

Fig. 2; pictures of last stage of surgical procedure and changed anterior fossae
a; showing after removing lesser and major wings of sphenoid bone and sphenoidal ridge
b; a picture after removing calvarium and the orbital bar
c; a orbital bar and pieces of bone put back on the frontal lobes and superior sagittal sinus
d; small anterior fossae before surgery
e; these were markedly enlarged after surgery

Fig. 3; chronological changes in DQ in 136 post-operative patients

Fig. 4; case one (a moderate trigonocephaly)
a; keep walking around in the room before surgery. He could say any meaningful words only keep saying “da, da, da”
b; radiological studies showed marked digital markings on the plain skull X-Ray, metopic ridge and small anterior fossae on 3D-CT
c; ICP monitoring during surgery showing high ICP in

normocapnea

d; reading cards like “water melon” “pillow” etc. with his father and playing rope skipping with his mother. He is saying ouch after hitting his shoulder to the wall at four months after surgery

Fig. 5; second case (a milder trigonocephaly)

a; radiological studies showed no digital markings on the skull X-Ray, shallower metopic ridge, broader forehead and bigger anterior fossae for mild trigonocephaly on 3D-CT and almost normal size of frontal lobes on MRI

b; ICP monitoring during surgery showing high ICP in normocapnea