

Nursing Care on Improving Postoperative Condition in CIPA Patients Combined with Charcot Spine

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Research

Keywords: Congenital insensitivity to pain with anhidrosis (CIPA), Charcot spine, Nursing care, Temperature monitoring, Posture management

Posted Date: January 5th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-139013/v1>

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Abstract

Background

Congenital insensitivity to pain with anhidrosis (CIPA) is a rare, hereditary, sensory autonomic neuropathy. There are few reports on CIPA combined with Charcot spine. Operation is a most effective method for CIPA patients who combine with Charcot spine, but those patients are easy to get hyperthermia and internal fixation failure after operation. Patients' deficit teeth also make nutrition management difficult. In order to help patients to avoid these problems, we designed a series of preoperative and postoperative nursing means: 1. Rectal temperature monitoring and Temperature control 2. Use new posture management 3. Assess the nutritional status of patients and make a diet plan. In this study, we aimed to evaluate the effect of these nursing means.

Methods

We retrospectively analysed the records of 3 CIPA patients combined with Charcot spine who accepted our nursing means to examine the effect of the nursing means.

Results

Rectal thermometry was more accurate than axillary thermometry. Physical cooling prevented hyperthermia for CIPA patients after operation. None of 3 patients had internal fixation failure. The BMI of one patient was only 14.0(malnutrition) before surgery, through nutritional intervention, all 3 patients' serum albumin levels returned to normal before discharge from the hospital.

Conclusions

In summary, this study has made a lot of suggestions that can improve the prognosis of CIPA patients with Charcot spine.

Background

Congenital insensitivity to pain with anhidrosis (CIPA) is a rare, hereditary, sensory autonomic neuropathy, characterized by absence of reaction to painful stimuli[1], inability to sweat and mental retardation[2, 3], it also can induce the destruction of the joint - neuropathic arthropathy (Charcot joint, Charcot spine). There are few reports associated with CIPA combined with the Charcot spine[4], and there is no standard treatment[5, 6]. CIPA is a really dangerous disease, and most patients cannot live over 25 years[7]. Operation is a most effective and reasonable method for those patients combined; however, there are also many challenges for patients to have the operation. For example, besides sensory autonomic neuropathy, patients are also insensitive to pharmaceutical cooling[8], they are easy to get hyperthermia or convulsion, or they can die of secondary infection caused by hyperthermia. The absence of reaction to painful stimuli is easy to cause internal fixation failure due to hypermobility. CIPA patients bleed more in 360 ° fusion operation, and they also have deficit teeth, which makes nutrition management difficult. We designed a series of nursing means to improve the prognosis of CIPA patients. The goals of this study are as follows: 1. to evaluate the effect of body position management. 2. to evaluate the effect of body temperature monitoring and control. 3. to assess the nutritional status of patients and make a diet plan.

Methods

Three CIPA patients with Charcot spine who received surgeries in our hospital from January 2010 to September 2019 were studied. In these 3 patients, besides lumbar vertebral injuries, CIPA can also do damage to the ankle, elbow, knee, and hip joint. The general information of the patients is shown in Table 1. The Mean operation age of these patients was $19.67 \pm$

5.03 (15–25). The average length of stay (ALOS) was 24.67 ± 6.03 days (19–31 days). The Median follow-up time was 14 months (6-124 months).

Table 1
General information of the patients with CIPA

Case	Age	Affected area	Operative name	Operation time (min)	Blood loss (ml)	Blood transfusion (ml)	Length of stay (d)	Follow-up (month)	Total drainage volume (ml)
1	15	Lumbar, ankles	Posterior osteotomy (L2-3), kyphosis correction, excision of intraspinal mass, internal fixation, Pyramesh vertebral reconstruction, autogenous bone graft fusion	345	1200	RBC 8u FFP 800	31	124	700 POD2
2	25	Lumbar, left elbow, left hip, left knee, left ankle	Right posterior iliac bone extraction, posterior vertebral body partial resection (L3, L4), interbody fusion (L2 / 3, L4 / 5, L5 / S1), spinal canal decompression(L2-S1), internal fixation, bone graft fusion	480	1200	RBC 4u FFP 400	24	14	850 POD5
3	19	Lumbar spine, left elbow, left knee, right ankle	Posterior left iliac bone extraction, part of L1 and L3 vertebral bodies and L2 vertebral body osteotomy, osteotomy space bone graft, pedicle screw fixation(T10-S1), interbody fusion (L4-5, L5-S1), bone graft fusion (L4-5, L5-S1, T10-S1)	385	1500	ABT 750 RBC 5u FFP 600	19	6	1365 POD7
POD, Post-operative day; RBC, Red blood cell; ABT, Autologous blood transfusion; FFP, Fresh frozen plasma.									

The nursing measures is described in Table 2.

Table 2
Nursing measures

Routine care	Specific nursing
<p>Preoperative examination and evaluation</p> <p>Physical examination on admission</p> <p>Post admission examination</p>	<p>Body position</p> <p>Bed rest: do preoperative evaluation of spinal cord compression, have bed rest if necessary</p> <p>Body position change: teach patients the method of bed and chair transfer, lying down → sitting up and sitting up → lying down</p> <p>Standing: wear braces until internal fixation fusion is completely stable; use tripeptide to accelerate bone healing</p>
<p>Temperature monitoring and prevention of hyperthermia</p> <p>Preoperative: monitoring axillary temperature and evaluating patients' tolerance to postoperative actual temperature monitoring</p> <p>Intraoperative: actual and nasal temperature monitoring</p> <p>Postoperative: actual temperature and axillary temperature (Figure) monitoring</p>	<p>Nerve function monitoring</p> <p>Intraoperative spinal cord monitoring</p> <p>Dynamic monitoring of sensation, movement, urine and stool</p>
<p>Psychology</p> <p>Encourage patients to express</p> <p>Carefully observe the abnormal physical signs and behaviors of patients, find out the causes, and do nursing intervention</p>	<p>Wound drainage</p> <p>Observe the volume of drainage to ensure that the drainage is sufficient. Avoid hyperthermia caused by heat absorption</p>
<p>Gastrointestinal management (diet, excretion)</p> <p>Assess the nutritional status of patients and make a diet plan (Figure)</p> <p>Promote the recovery of gastrointestinal function after operation</p>	<p>Function exercise</p> <p>During bedtime</p> <p>Underground activities</p>
<p>Blood volume management</p> <p>Monitor heart rate and blood pressure, keep blood volume stable</p> <p>Follow the doctor's advice to use vasopressor drug if necessary</p>	

The main points of nursing 1: Temperature monitoring and Prevention of hyperthermia.

Temperature monitoring:

Rectal thermometry is recognized as the most accurate surrogate of body temperature, especially for hyperthermia patients[9]. We decided to monitor rectal temperature for these patients, compared the result of rectal temperature with axillary temperature. For rectal temperature monitoring, common ECG monitors (including ECG monitors of general company, Mindray Medical International, Philips, Siemens, and GE) can be used. It is necessary to prepare the temperature probe conversion line, which is suitable for the TEMP probe interface of the monitor in advance. A probe needs to be inserted into the rectum via the anus and kept in the patient's rectum. In order to ensure patients can tolerate and cooperate with rectal temperature monitoring after the operation: do a pre-operative rectal temperature monitoring for

patients to assess their compliance and increase the likelihood of their postoperative cooperation. Measurement method: lubricate the probe and then put it into the intestinal cavity according to the clinical procedure. Caution: If resistance is felt while inserting the probe, stop the insertion and pull back the probe slightly. Do not force the probe in because improper placement may damage the patient's cavity mucosa. The probe has a scale, which can be used to determine the depth of probe placement and see whether the probe is falling off or retreating through the exposed scale.

Measure axillary temperature at 2 pm every day before surgery. Measure the rectal temperature and axillary temperature at 6 pm-6 am-2 pm-6 pm every day since one day prior to the operation, to identify hyperthermia in time.

Temperature control

Prepare temperature and humidity instruments for patients, because they can't reduce the temperature by perspiration. Maintain the ambient temperature at 23 ± 2 °C. Reduce the patient's cover at the same time. The ice bag is used for physical cooling to avoid hyperthermia after the operation. If the rectal temperature is maintained between 37.5 to 38.5°C, physical cooling is the main way to prevent hyperthermia or convulsion. If the rectal temperature is higher than 38.5 °C and the drop in body temperature is not substantial after physical cooling, it is necessary to use aspirin/lysine to lower the hypothalamus temperature set-point, despite the fact that CIPA patients are relatively insensitive to drug cooling.

The main points of nursing 2: Posture management

Bed rest

Preoperative: evaluate the degree of spinal cord compression by imaging and determine whether patients should have bed rest before surgery. One patient in this study had spinal cord compression before surgery, so he was required to lie in bed before the operation. Due to the mental retardation and stubborn character of CIPA patients, he did not understand why he needed to lie in bed before the operation. He insisted on getting up and going to the bathroom. Family members could not fully understand the importance of bed rest after the communication and they were required to accompany the patient 24/7. Family members worked with the medical staff to ensure that patient lies in bed before the operation. Avoid further compression of the spinal cord or paralysis caused by activities.

Postoperative: Because of the absence of reaction to painful stimuli, the patient's body position cannot be limited by pain. Teaching patients how to turn over and sit up can avoid the failure of internal fixation caused by excessive activity.

Position change

The postoperative bedtime is relatively long because of the osteoporosis and operation. An X-ray should be taken before the patients get out of bed to make sure that the internal fixation is stable.

From lying position to standing position

Before going down to the ground, patients should put on the brace in bed first, and then sit up beside the bed to avoid internal fixation loosening or even displacement as their body position changes (see Fig. 1). They should not directly sit up from the supine position. Because at this time the bending force is too large, which can easily cause the loosening of internal fixation.

From standing position to lying position

when the patients lie down from sitting up, they should be wearing the brace. The brace can be removed after lying down. (see Fig. 2). During the whole process of sitting up and lying down, the brace can disperse the stress, reduce the pressure of internal fixation, and minimize the risk of internal fixation loosening.

Underground activities

Denervation of the bone can affect the healing. Teriparatide is used to promote the bone formation after the operation. Patients need to come back to the hospital for examination at the first, third, and sixth months after the operation. The results of the X-ray can be used to determine the time when the brace can be removed. Before the internal fixation is completely stable, the brace should be worn when changing the body position.

The main points of nursing 3: Assess the nutritional status of patients and make a diet plan

Nrs2002 is used to evaluate the status of patients. CIPA patients always have tooth problems[5, 10, 11]. Therefore, evaluation of patients' teeth and chewing ability should be done before making the intervention plan. (Fig. 3)

Data Collection

These patients were followed up by phone, and their outpatient or medical records were reviewed.

Results

Temperature

The changing trend of 3 patient's postoperative rectal and axillary temperature (Fig. 4).

Nutritional status

the BMI of one patient was only 14.0(malnutrition) before surgery. Through nutritional intervention, all 3 patients' serum albumin levels returned to normal before discharge from the hospital(Figure 5).

Discussion

1. CIPA is a rare hereditary sensory autonomic neuropathy. It was first reported by Swanson in 1963[12]. There were few reports of CIPA in the previous literature. CIPA is more common in men, and all three patients in this study are female. Dyck named these diseases as Hereditary sensory and autonomic neuropathy (HSAN)[13]. HSAN has been categorized into 5 types: HSAN type I (Hereditary sensory radicular neuropathy), HSAN type II (Congenital sensory neuropathy), HSAN type III (Familial dysautonomia), HSAN Type IV (CIPA), and HSAN type V (Congenital insensitivity to pain with partial anhidrosis)[2, 14]. The three cases reported in this study belong to type IV (CIPA).

2. The diagnosis of CIPA is based on pain, temperature test and iodine starch sweating qualitative test[15]. Further diagnosis requires molecular biotechnology to find out the location of gene mutation. In this study, all 3 cases are investigated by the above methods, and skin biopsies were performed in another hospital, which showed a loss of unmyelinated fibers of peripheral nerve, denervation of skin nerve and abnormality of exocrine gland. All the evidence supports the CIPA diagnosis.

3. Rectal thermometry is recognized as the most accurate surrogate of body temperature, especially for hyperthermia patients[9]. CIPA has a great impact on body temperature, so it is necessary to monitor the temperature of patients after operation. The axillary temperature is not accurate when the core body temperature is abnormal. We must pay attention to prevent death due to secondary infection caused by hyperthermia.

CIPA, as a rare sensory autonomic neuropathy, affects sweating and heat dissipation. In this study, we first tried to use rectal and axillary temperature to reflect the trend of postoperative temperature change. Secondly, we tried to explain the cause of the temperature change. The time course of temperature showed two peaks, on the first day and 5–7 days after surgery, respectively. The changing trend of rectal and axillary temperature is similar, but the sensitivity of axillary temperature is not as good as that of the rectal temperature. Axillary temperature is easy to cause false-negative results, especially in the early postoperative period. It is suggested that rectal temperature should be used to reflect the temperature changes, especially in the early postoperative period.

The reasons for two peaks: the peak on the first day after surgery is considered to be related to surgery; the second peak on the 5–7 days after surgery is considered to be a fever following the removal of the drainage tube. CIPA patients' bone healing is slow and poor [14, 16, 17], so they always get out of bed later than the regular patients. Before the removal of the drainage tube, the patients have to lay in the bed all the time. The surgical trauma is substantial, and there is much bleeding. After the removal of the drainage tube, there will be blood or fluid accumulation in the body, which leads to the second peak.

4. CIPA is similar to spinal cord injuries, especially the cervical spinal cord injury, because they all have sensory autonomic neuropathy, which can affect sweating and heat dissipation. Therefore, our study uses physical cooling to prevent hyperthermia caused by spinal cord injury, and it has achieved good results. Although CIPA patients are insensitive to drug cooling, we can still use aspirin/lysine to reduce their temperature by lowering the temperature set-point. It is confirmed that to some extent the method of CIPA postoperative temperature control can be applied the treatment of spinal cord injury.

5. CIPA combined with Charcot spine should have 360 °fusion, postoperative position management is very important. Vialle reported 9 cases of CIPA with Charcot spine and emphasized the importance of 360 °fusion. He pointed out that due to the lack of pain protection mechanism, the repeated excessive activity of patients is easy to cause the failure of internal fixation even in case of a simple posterior fusion [18]. Therefore 360 °fusion was performed in all 3 CIPA patients. 360 °fusion can provide the most powerful stability to prevent the internal fixation failure caused by excessive activity. 360 °fusion is important, but postoperative position management is also important. The position management is particularly important in the early postoperative period, especially in the period before the internal fusion is completely stable. Our method that ensure patients always wear braces during the whole process from lying position to standing position and from standing to lying position can disperse the stress of spine.

6. The BMI of one patient was only 14.0(malnutrition) before surgery. Through our nutritional intervention, this patient returned to normal before discharge from the hospital. This result can confirm that our approach can really help patients.

Declarations

Acknowledgements:

We acknowledge all of the members of Department of Orthopedic Surgery, Peking Union Medical College Hospital for helpful support and the patients for participating in this study.

Authors' contributions:

All authors read and approved the final manuscript. NG and YCP designed this study, participated in the workshop, and supervised the manuscript. BDT and ZYZ coordinated the study, analyzed data, and wrote the final manuscript. YT and SYC collected and interpreted data. JGZ participated in study design and assisted in the collection of data. NW participated in the workshop and collaborated in the feasibility study and reviewed the manuscript.

Funding:

No funding received.

Availability of data and materials:

The data analyzed during this study were obtained with the permission from the Peking Union Medical College Hospital from the corresponding author on reasonable request.

Ethics approval and consent to participate:

This study has been approved to the ethics committee at Peking Union Medical College Hospital.

Consent for publication

Consent for publication was obtained from all participants.

Competing interests

No competing interest.

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Figures

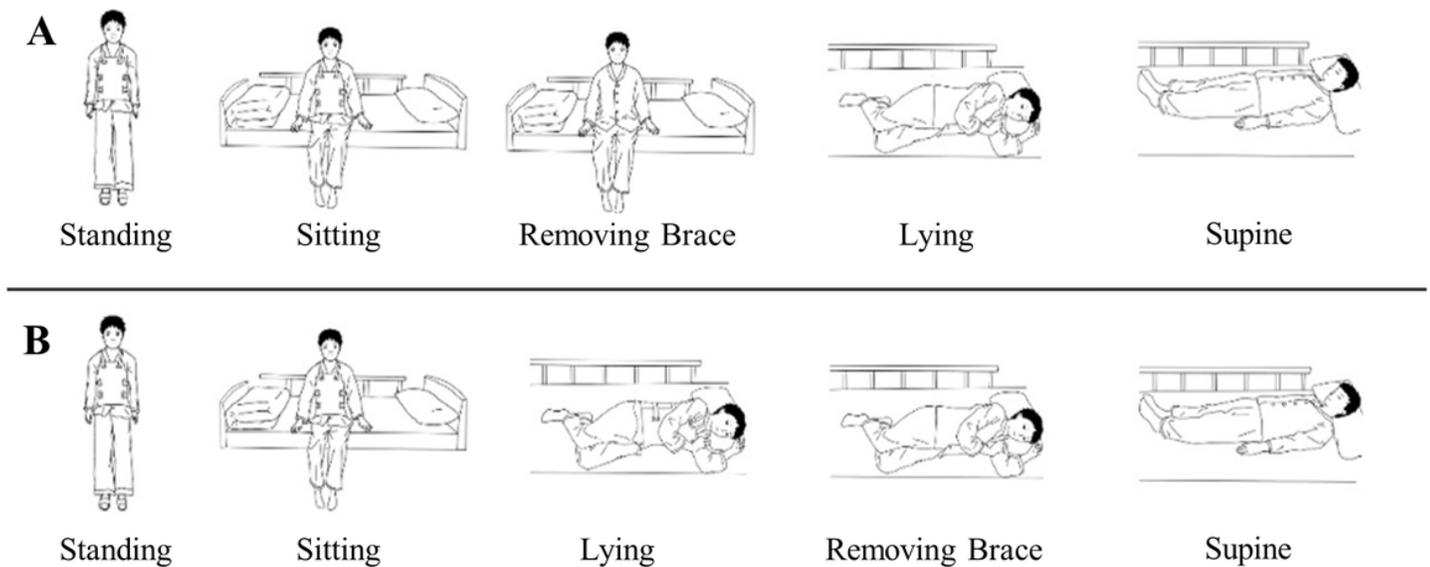


Figure 1

The comparison of postoperative normal spinal disease patient and CIPA patient from lying to standing ; A normal spinal disease patient; B CIPA patient

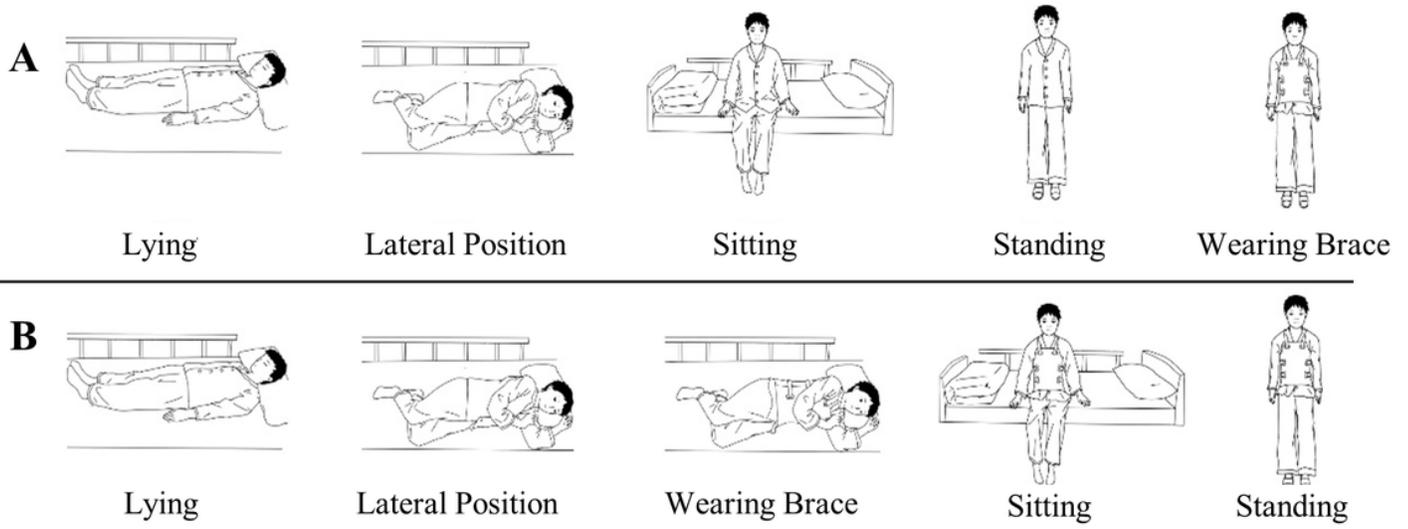


Figure 2

The comparison of postoperative normal spinal disease patient CIPA patient from standing to lying ; A normal spinal disease patient; B CIPA patient

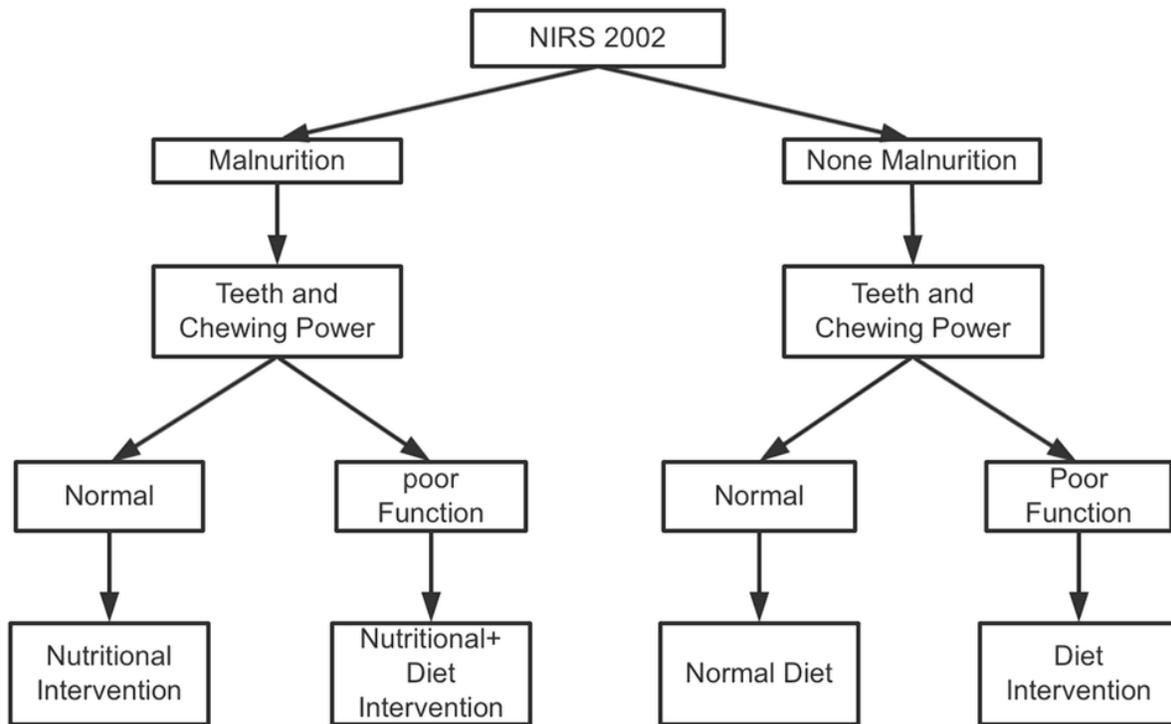
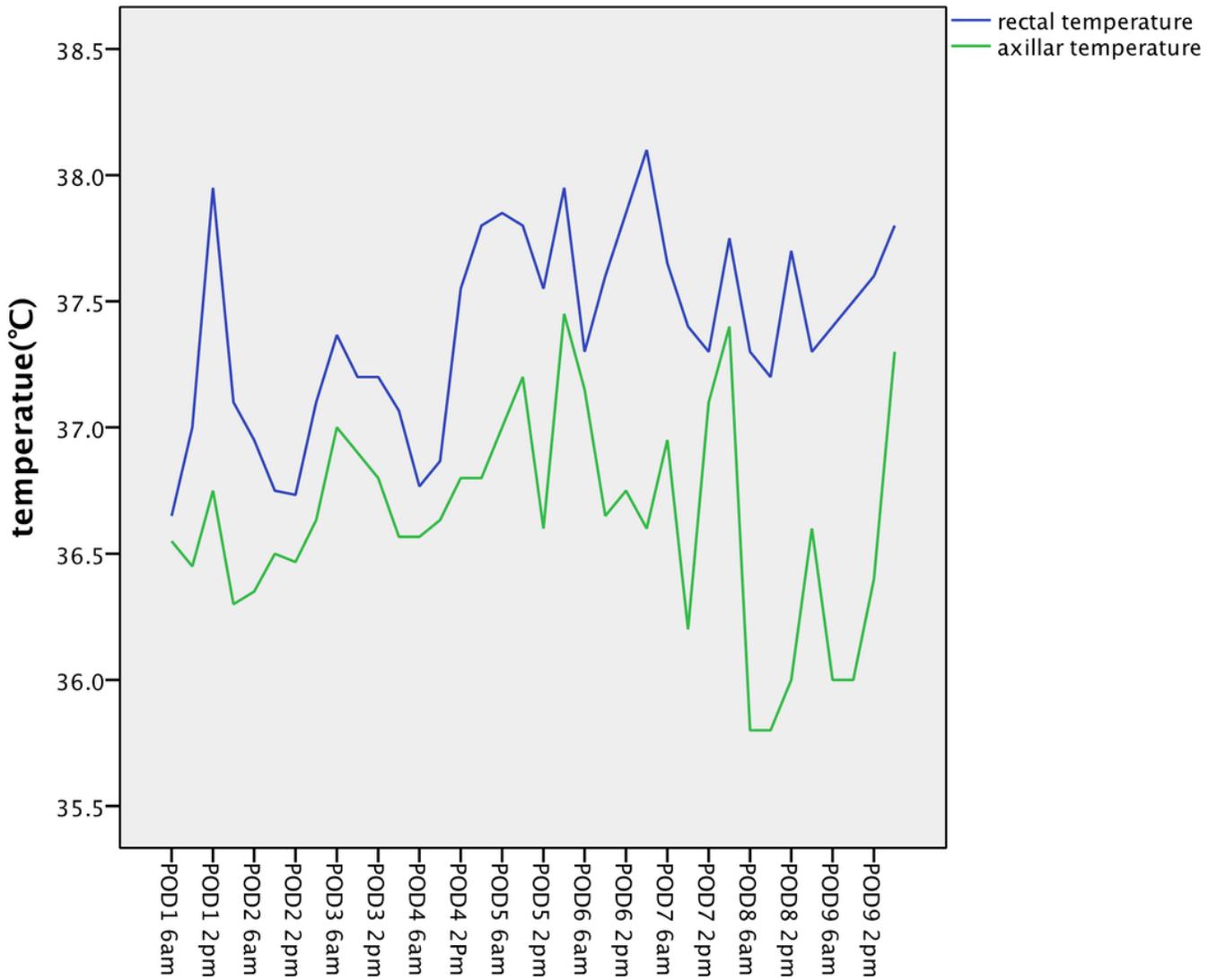


Figure 3

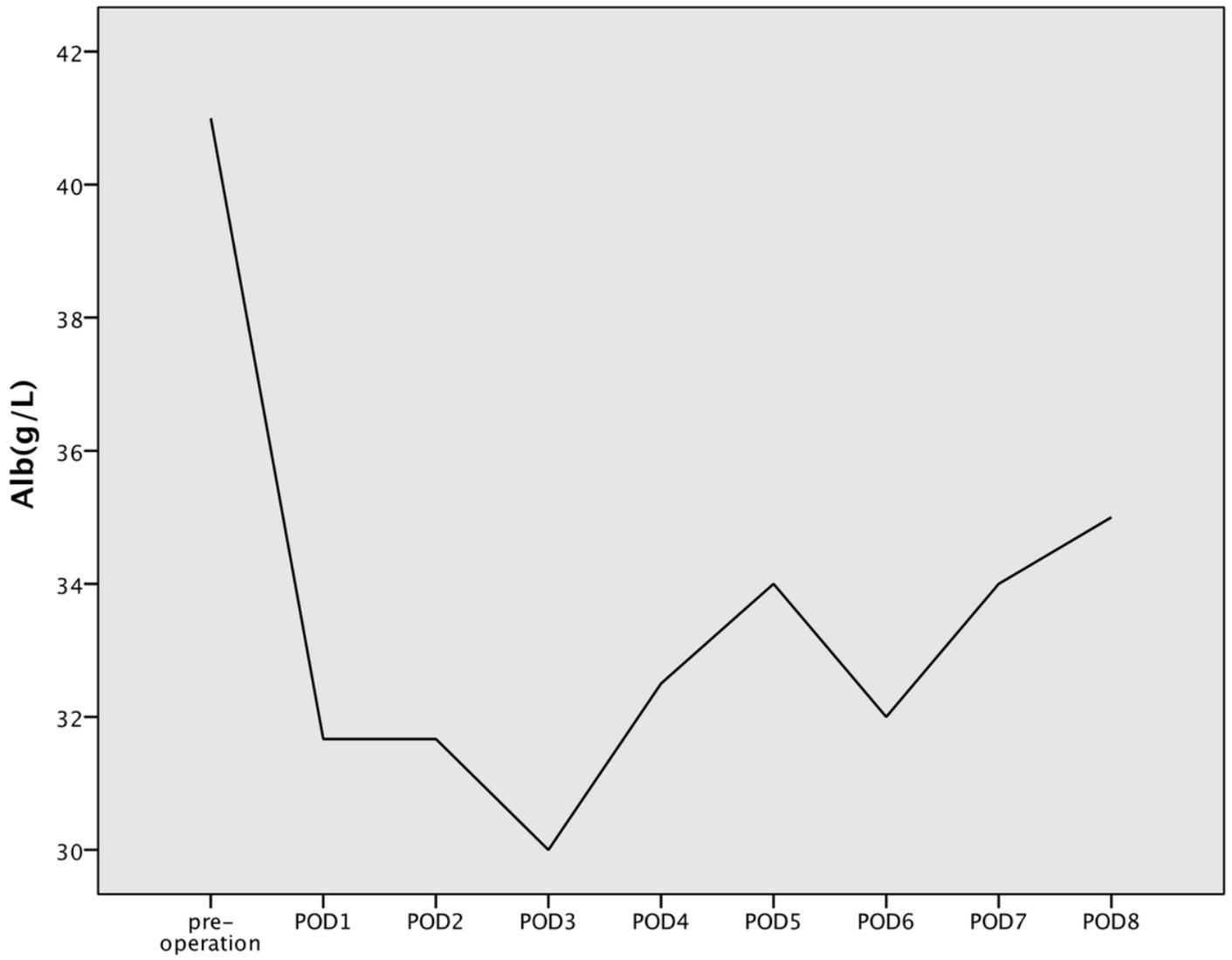
evaluation of patients' teeth and chewing ability



POD: Post-operative day

Figure 4

The change trend of 3 patient's postoperative rectal and axillary temperature



POD: Post-operative day

Figure 5

The nutritional status of 3 patient