

小児の運動生疾患の介護等に関する研究班

先天性無痛無汗症児の歯列成長発育

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研究要旨

先天性無痛無汗症の歯科的な問題として、舌や口唇、頬粘膜の咬傷、手指の咬傷、う蝕や歯周疾患に起因した上下顎骨髄炎、外傷による裂傷や顎骨骨折などが挙げられる。そこで咬傷および歯の早期喪失の予防のため保護プレートを着用し良好な結果を得ているが、長期にわたる保護プレートの使用で歯列、歯槽部の発育を抑制してしまう可能性があると考えられる。これについて生後 10 カ月から保護プレートを使用している女児（症例 1）および乳幼児期に下顎前歯を抜歯された男児 2 例（症例 2, 3）について歯列模型計測を行い検討した。

症例 1 では上下各歯牙間幅径は 4 才 9 カ月時、下顎乳犬歯部-1.1SD の他は著明に小さい部は認められなかった。症例 3 は永久歯列で上下顎共著明な前歯部の叢生が認められたが、下顎乳前歯 6 本早期に抜歯された既往があり、早期に多数歯を喪失することにより歯槽部の成長に影響した可能性があると考えられた。

A. 研究目的

先天性無痛無汗症の歯科的な問題として、舌や口唇、頬粘膜の咬傷、手指の咬傷、う蝕や歯周疾患に起因した上下顎骨髄炎、外傷による裂傷や顎骨骨折などが挙げられる。特に生後 6 カ月ごろから最初の乳歯が萌出するが、それに合わせて舌や口唇、頬粘膜の咬傷が始まり、このような咬傷は本症のすべての患者に認められている。そこでこれらに対し、保護プレートを作製し装着させ、咬傷および歯の早期喪失の予防を図り良好な結果を得ている。しかしプレートの問題点として、破損および歯の萌出に伴いプレートが合わなくなってくるため、度重なる再作製が必要であること、また長期にわたるプレートの使用で歯列、歯槽部の発育を抑制してしまう可能性があることなどが挙げられる。今回生後 10 カ月から保護プレートを使用している女児（症

例 1）の歯列の成長発育について歯列模型を用い経年的に計測を行うと同時に、乳幼児期に下顎前歯を抜歯された男児 2 例（症例 2, 3）についても歯列模型計測を行い検討した。

B. 研究方法

図 1 に模型計測部位を示す。また永久歯列については歯列弓幅径は第 1 小臼歯頬側咬頭頂間距離を、歯列弓長径は両側中切歯唇面接線から両側第 1 大臼歯遠心面を結んだ線までの距離を計測した。

C. 研究結果および考察

症例 1 の計測値は表 1 に示す。上下顎各歯牙間幅径は上顎第 1 乳臼歯部を除いて 4 才 1 カ月まで漸次増加を示し、特に下顎第 2 乳臼歯部は 3 才 5 カ月から 4 才 1 カ月の間に急激に増加が認められた。また 4 才 9 カ月時には下顎乳犬歯部-1.1SD の他は歯列弓幅径の著しく小さい部は認められない。尚、下顎左側

乳中切歯は3才8カ月時脱落している。歯列弓長径については、上顎 A-E 間および下顎 A-E、A-E_D 間は漸次減少しているが、これは第2乳臼歯が近心移動したためと思われた。また下顎では4才1カ月時いずれの計測部位も大きく減少していた。これは3才8カ月時下顎左側乳中切歯が脱落したことによる。前歯部の排列が変化した結果であると思われる。このように歯を失うことが、歯列の形態に大きな影響を与えることが示唆された。

歯列の成長については2才7カ月から4才9カ月までの成長量を比較できる対象がなく、プレートによる抑制については明らかに影響を受けているかについて、歯の傾斜度、歯槽部の形態、正貌および側貌頭部X線規格写真など他の方法での検討が必要である。症例1は4才3カ月頃より咬傷が少なくなり、現在プレートの使用時間を徐々に減らしているところである。

症例2の計測値は表1 No2 に示す。生後2週間で無痛無汗症と診断された男児の3才時の歯列模型である。生後6カ月で下顎乳中切歯萌出するもすぐに著しい動揺、咬傷のため、早期に抜歯されている。その後下顎右側第1乳臼歯から下顎左側乳犬歯まで萌出後すぐに抜歯されている。上顎歯列幅径乳犬歯舌側歯頸最下点間距離が-2,3SD の他は-1SD 以内であった。また下顎歯列弓幅径第2乳臼歯部については1SD 以内で特に下顎臼歯部の狭窄は認められなかった。

症例3は2才5カ月時無痛無汗症と診断された男児の15才時の歯列模型である。上顎では永久歯が並びきらず、左右第1小白歯を抜歯しているが、V字歯列弓で前歯部叢生、左右中切歯の著明な唇側転位を認める。下顎では右側中切歯抜歯、左側側切歯先天性欠如

で2歯欠損しているにもかかわらず、前歯部の叢生、著明な右側側切歯の舌側転位が認められる。歯冠幅径については表2に示すように上下顎とも標準的かやや小さく、下顎第2小白歯は-2SD 以上小さい歯冠幅径であった。患児は1才半の時にはすでに下顎前歯6本早期に抜歯されており、早期に多数歯を喪失することにより歯槽部の成長に影響した可能性があると推測された。

D. 結論

症例1では4才3カ月頃より咬傷が少なくなりプレートの使用時間を徐々に減らしているが、4才9カ月時、上下各歯牙間幅径は、下顎乳犬歯部-1.1SD の他は著明に小さい部は認められなかった。症例3は永久歯列で上下顎共著明な前歯部の叢生が認められたが、下顎乳前歯6本早期に抜歯された既往があり、早期に多数歯を喪失することにより歯槽部の成長に影響した可能性があると推測された。

正常咬合を有する小児の上顎および下顎歯牙間幅径は漸次増加を示す。このような歯列の成長のためにはプレートを使用せず、自然な成長を発現できるようにしたいが、乳幼児期の咬傷の著しい時期、プレートにより咬傷を予防し、さらに早期に歯を失うことによる咀嚼能率の低下を防止することも無痛無汗症児には必要であり、この時期成長抑制傾向であったにせよ咬傷の減少とともにプレートははずすことが可能となり、その後歯列の成長が catch up するようであればプレート使用を選択してよいと思われる。

E. 参考文献

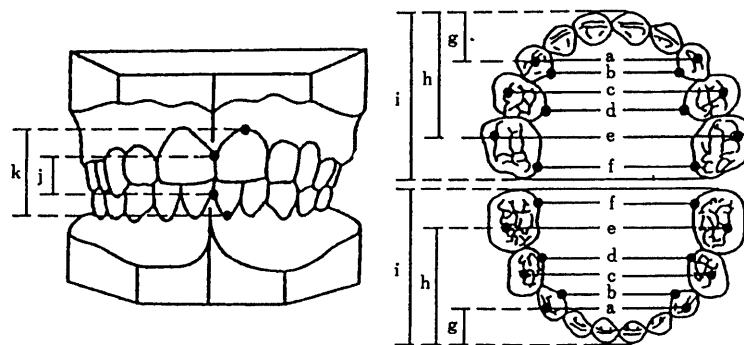
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図1 計測部位
(乳歯列)



歯列弓幅径

- a : 上下顎両側犬歯咬頭頂間距離 (C_c-C_c)
- b : 上下顎両側犬歯口蓋(舌)側歯頸部最下点間距離 (C_L-C_L)
- c : 上顎両側第一乳臼歯頬側咬頭頂間距離 (D-D)
- 下顎両側第一乳臼歯頬側分界溝間距離 (D-D)
- d : 上顎両側第一乳臼歯口蓋側歯頸部最下点間距離 (D_L-D_L)
- 下顎両側第一乳臼歯舌側近遠心咬頭頂間の舌側溝直下点間距離 (D_L-D_L)
- e : 上顎両側第二乳臼歯頬側分界溝間距離 (E-E)
- 下顎両側第二乳臼歯近心頬側分界溝間距離 (E-E)
- f : 上下顎両側第二乳臼歯口蓋(舌)側歯頸部最下点間距離 (E_L-E_L)

歯列弓長径

- g : 両側乳中切歯の唇面を連ねた線の中央から垂線を降ろした
両側乳犬歯咬頭頂を結んだ線までの距離 (A-C_c)
- h : 〃
両側第二乳臼歯近心頬側分界溝間を結んだ線までの距離 (A-E)
- i : 〃
両側第二乳臼歯最遠心端間を結んだ線までの距離 (A-E_D)

歯列弓高径 (咬合平面を基準として)

- j : 上下顎両側乳中切歯間の歯間乳頭間距離 (Dental Height)
- k : 上下顎左側乳中切歯唇面歯頸部中央間距離 (ULA-LLA)

表 1 ANALYSIS OF DENTITION

							mm(SD)
		2Y7M	3Y0M	No 1 3Y5M	4Y1M	4Y9M	No 2 3Y0M
ARCH WIDTH Maxilla	Cc-Cc	27.55(-1.5)	27.95(-1.0)	28.20(-0.9)	28.95(-0.3)	29.00(-0.2)	29.00(-0.9)
	C _L -C _L	29.55(4.5)	22.75(-0.9)	22.55(-1.1)	22.55(-1.1)	23.75(-0.1)	21.80(-2.3)
	D - D	37.10(-0.2)	36.30(-0.6)	37.40(-0.1)	37.40(-0.1)	37.40(-0.1)	39.55(0.0)
	D _L -D _L	26.00(-0.4)	25.00(-0.9)	25.40(-0.7)	25.80(-0.5)	25.70(-0.5)	26.70(-0.6)
	E - E	43.30(-1.0)	43.95(-0.6)	44.30(-0.4)	44.30(-0.4)	45.25(0.2)	44.60(-1.0)
	E _L -E _L	31.00(1.6)	28.70(0.0)	28.50(-0.1)	28.75(0.0)	28.75(0.0)	29.55(-0.3)
ARCH WIDTH Mandible	Cc-Cc	20.65(-1.7)	20.75(-1.6)	21.00(-1.4)	21.50(-1.0)	21.35(-1.1)	
	C _L -C _L	17.60(-0.6)	16.50(-1.5)	17.90(-0.3)	17.65(-0.5)	17.00(-1.1)	
	D - D	32.00(-0.1)	32.50(0.3)	32.50(0.3)	33.60(1.0)	32.75(0.5)	
	D _L -D _L	24.15(0.0)	23.75(-0.3)	23.85(-0.2)	24.55(0.3)	24.30(0.1)	
	E - E	38.95(0.8)	39.65(1.3)	39.70(1.3)	41.80(2.8)	41.25(2.4)	39.55(0.3)
	E _L -E _L	28.25(0.5)	28.50(0.7)	28.60(0.7)	30.20(1.9)	29.55(1.4)	29.30(0.4)
ARCH LENGTH Maxilla	A -Cc	9.40(1.5)	9.45(1.6)	9.55(1.7)	8.80(0.8)	7.30(-0.8)	
	A - E	23.35(0.6)	23.10(0.4)	23.10(0.4)	22.40(-0.1)	21.00(-1.2)	
	A -E _D	-	-	-	-	-	
ARCH LENGTH Mandible	A -Cc	4.30(-1.1)	4.40(-1.0)	4.60(-0.7)	3.15(-2.5)	2.20(-3.6)	
	A - E	18.80(-0.3)	18.25(-0.8)	18.00(-1.1)	17.00(-2.0)	15.50(-3.3)	
	A -E _D	24.20(-1.0)	23.85(-1.3)	23.75(-1.3)	22.60(-2.3)	21.80(-3.0)	

		No 3 15Y0M
ARCH WIDTH	Maxilla	36.10(-3.3)
	Mandible	26.00(-5.2)
ARCH LENGTH	Maxilla	31.80(-1.9)
	Mandible	25.00(-3.3)

表 2 MESIODISTAL CROWN DIAMETERS

		mm(SD)			
LOCATE		No 1 (F)		No 2 (M)	
		R	L	R	L
Maxilla	A	6.30(-0.5)	6.20(-0.8)	-	-
	B	5.15(-0.5)	5.15(-0.5)	-	-
	C	6.80(0.8)	6.95(1.2)	6.60(-0.2)	6.45(-0.5)
	D	7.25(0.2)	6.75(-1.1)	7.55(0.5)	7.75(1.0)
	E	-	-	9.25(-0.1)	9.30(0.0)
Mandible	A	3.90(-0.7)	3.60(-1.8)	-	-
	B	-	4.60(-0.2)	-	-
	C	7.00 *	5.70(-0.1)	-	-
	D	8.50(0.9)	8.20(0.3)	8.30(0.1)	-
	E	10.10(0.0)	9.80(-0.6)	10.60(0.5)	10.80(0.9)

*癒合歯

LOCATE		No 3 (M)	
		R	L
Maxilla	1	8.15(-0.8)	8.15(-0.8)
	2	6.40(-0.9)	6.30(-1.0)
	3	7.95(-0.2)	7.95(-0.2)
	4	-	-
	5	6.40(-1.2)	6.25(-1.5)
	6	10.25(-1.2)	10.40(-0.9)
Mandible	1	-	5.60(0.4)
	2	5.80(-0.4)	-
	3	6.80(-0.7)	7.00(-0.3)
	4	6.20(-2.4)	6.30(-2.1)
	5	6.90(-0.5)	7.10(0.0)
	6	11.40(0.0)	11.40(0.0)

**DENTAL GUIDLINES OF HEREDITARY
SENSORY AND AUTONOMIC NEUROPATHY WITH ANHIDROSIS
(HSAN)**

MINISTRY OF HEALTH AND WELFARE :Study on System of Good Rearing of Children
with High Risk; *Study on Case of Children with Motor Disorders*

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INTRODUCTION

Sensory neuropathy with anhidrosis is a hereditary disease characterized mainly by insensitivity to pain, anhidrosis (an abnormal deficiency of sweat), and mental retardation. It is said that this disorder was first reported by Nishida et al. in 1951. According to current classification system, hereditary peripheral neuropathies are divided into hereditary motor and sensory neuropathy and hereditary sensory and autonomic neuropathy. Thus the disorder comes under the hereditary sensory and autonomic neuropathy type IV. Thus far there have been around 100 cases in Japan. Recently, Iendo et al. reported that the lack of the nerve growth factor (NGF) on chromosome 1 causes this disease.

The insensitivity to pain is derived from the lack of thin myelinated and unmyelinated nerve fibers which conduct pain impulses. The sweat deficiency results from the lack of the peripheral sympathetic end-fibers which innervate blood vessels surrounding the sweat glands.

The clinical complications resulting from the lack of pain include tongue and mouth injuries, fractures, osteomyelitis, Charcot's joint, skin injuries, and burns. Unfortunately these complications are common and often severe. The complication associated with anhidrosis is the disturbed regulation of body temperature. Further, most patients with this disease manifest convulsive seizures and mental retardation. This suggests that the central nervous system may be involved in this disorder. Although the insensitivity to pain is one of the characteristic symptoms, the other sensations are not disturbed.

Oral Manifestations

The problems in this disorder include bite wounds on the tongue, lips, cheek mucosa, and finger tips, osteomyelitis of the upper and lower jaws due to dental caries and/or periodontal infection, lacerations and fractures caused by trauma, etc. Concurrently with deciduous teeth eruption starting around 6 months after birth, bite wounds on the tongue, lips, and cheek mucosa are first found, at this time the patient may be first diagnosed as this disease. These bite wounds are seen in almost every patient. In many cases the teeth will be extracted to prevent continuous bleeding from the tongue and/or lips. It is easily inferred that such early tooth loss will adversely affect the growth not only of the alveolar and jaw bones but of the body, because of deteriorated masticatory function and associated nutritional defects. If the teeth are conserved in spite of repeated bite wounds, the tongue will almost always be scarred, and the lingual papillae and taste buds will disappear, resulting in abnormal taste sensation. The clinical conditions of this disease and preventive measures will be described in this guidelines.

Dental problems by age

1) Infancy

Among milk teeth, the lower central incisors start to erupt around 6 months after birth. An ulcer may occur on the inferior surface of the tongue due to a congenital tooth in neonatal period even in the normal infant. This condition is called Riga-Fede's disease and is a sort of abrasion resulted from to-and-fro movements of the tongue. The causal tooth/teeth used to be extracted when the infant had fever or suckling disturbance, or hurt mother's nipples. Recently, however, conservative procedures are common for this trouble.

In the child with sensory neuropathy with anhidrosis, an ulcer occurs on the inferior surface of the tongue often with bleeding, which can penetrate almost to the muscular stratum. The upper deciduous incisors erupting around 10 months of age, with which the tongue can be bitten, sometimes completely severing the tip. Other than tongue biting, the patients may experience loosened or lost teeth resulting from severe bruxism or strong biting of toys, cords, etc., for the infant is commonly very eager to take anything in the mouth. In most cases, however, they may manipulate the teeth by themselves causing self-extraction probably because of an uncomfortable feeling with eruption. Bite wounds will involve the lateral edges and/or the dorsum of tongue or the buccal mucosa with the eruption of deciduous molars.

2) Early childhood

Primary dentition with 20 teeth becomes complete around 2 and one-half years of age. The most common injury is the tongue, although the patient may often bite the buccal mucosa, lips, and nails and the finger tips resulting in severe wounds. In this period, most patients have lost several teeth, and tongue thrusting can make the bite wounds more severe.

Dental caries may progress undetected because the patient does not feel pain. It seems possible that the patient could be unaware of his/her trouble even at the stage of abscess or cellulitis, without any subjective symptom during the stages of pulpitis and periodontitis. There has been one case of jaw fracture after mandibular osteomyelitis caused from caries infection, illustrating why the patient must regularly visit his/her dentist for the examination of oral condition, treatment and prophylaxis.

3) School period

The eruption of permanent teeth starts around 6 years of age, and the incidence of self-injuries by biting is lower than that in primary dentition. It is not clear if this is learned behaviour from the preschool dentition. It is not clear if this is learned behaviour from the preschool period. It is also possible the tongue edge has been so severely scarred and shortened that the patient can no longer bite the tongue as readily. However, bite wounds, self-extraction, and severe tooth attrition due to bruxism may be found and may be referable to the discomfort of milk tooth shedding and permanent tooth eruption. The patient should have regular dental surveillance, because he/she may not complain of pain even when suffering from pulp inflammation or infection. In addition, the patient should be supported mentally, for apprehension, fear, and conflict may result in behaviour that may increase self-extraction and bruxism.

4) Adolescence

During the adolescent period, prosthodontic treatment may be necessary both for the rehabilitation of masticatory function and for the prevention of cheek biting, for many patients have lost teeth by this time. The denture should be frequently examined and may need to be repaired or even remake as the jaws grow. The wisdom teeth erupt after 17 to 18 years of age. In particular, the lower third molar is susceptible to purulent pericoronitis due to the lack of space for eruption. The symptoms of pericoronitis include fever, swelling, foetor exore (mouth odor), trismus (disturbance of mouth opening) or dysphagia (swallowing disturbance).

Since not all persons will have four wisdom teeth, it is advisable that a panoramic radiograph is taken to detect the presence of third molar(s) and measure the space for eruption.

5) Adulthood

We have experienced not many adult patients with sensory neuropathy with anhidrosis, therefore the dental problems for adult is not well known. But it appears that there are fewer difficulties compared with the infant and child periods. Denture wearing is commonly recommended due to tooth missing as a result of caries, pericoronitis, trauma, or self-extraction. A fixed, non-removable denture (bridge) can be set with safety if the number of missing teeth is a few and the patient does not grind the teeth.

Removable dentures are indicated in cases with many missing teeth. Close attention should be paid to the oral condition of the patient, as he/she does not feel pain and can tolerate an ill-fitting denture resulting in ulcers, loosened teeth, and sometimes bone exposure.

Dental considerations

1) Prevention and treatment of bite wounds

A protecting plate should be applied to the patient when the tooth is erupting and a bite wound appears. Thermoplastic resin of 0.6 to 0.8 mm thick is useful in making the plate. When the patient frequently has bite wounds in the region of missing teeth or in the space for eruption, it can be effective to fill the space with soft resin. These devices must be remade according to tooth eruption and jaw growth.

2) Prevention and treatment of dental caries

For caries prevention plaque removal is important. Increase the times of taking foods and drinks with sugar (sweets restriction).

Also the use of fluoride painting and fluoride containing dentifrice is effective for strengthening teeth. Regular dental examinations are, of course, recommended and early treatment is important when dental caries are detected.

3) Prevention and treatment of periodontal disease

Both gingivitis (inflammation of the gum) and periodontitis (infection and destruction of the structure surrounding the tooth roots) are caused by the infection of obligate anaerobes such as *Spirochaeta* and *Porphyromonas*.

The prevention of periodontal disease consists of removal of plaque and calculus and periodontal treatment. Periodontal condition is also affected by fever and malnutrition.

4) Dental complications associated with epilepsy

Seventeen percent of children with sensory neuropathy with anhidrosis will also have epilepsy.

The dental complications with epilepsy include tooth injuries such as fracture and avulsion and jaw fracture. There may be no symptoms shortly after injury and the symptoms of these traumas may manifest after a few weeks or months later. The patient should have a dental examination when the history suggests orofacial injury.

Some antiepileptic drugs may provoke hyperplasia (increased growth) of the gum. One-half of patients taking phenytoin (diphenylhydantoin) manifest this gum hyperplasia which will attain a maximum size in adolescence in many cases. Gingivitis can increase the hyperplasia but this complication can be prevented by cleaning the mouth and tooth surfaces with tooth brushing.

Sometimes an aggressive dental management such as gingivectomy (a surgical removal of the enlarged gingival margin) may be necessary when there occurs a cosmetic and/or functional disturbance.

[Case 1]

Recurrent ulceration and bleeding occurred on the inferior surface of the tongue following the eruption of the lower front teeth in a 10th month old child. A protecting plate was immediately set in place, and the ulcer began to heal. After months (1 year of age), the patient bit off the apex of tongue as the upper front teeth erupted. A protecting plate was set on the upper jaw too. Bite wounds occurred on the lateral edges of tongue as the upper lateral incisors erupted at 14 months of age. The bite wounds of the tongue edge and cheek mucosa became more remarkable after the eruption of the upper and lower first deciduous molars at 1 and 1/2 years of age. After that, we presumed that abrasions from tongue thrusting to the space for eruption was one of the causes of trauma, so we changed to the protecting plate shown in Fig.19 and Fig.20 which was made to fill that space, after which the cheek wound healed. Similar plates were successfully applied until the eruption of the upper and lower second deciduous molars with good bite wound prevention. When the plates are in place a fluoride gel should be applied to the inside of the plate to help prevent caries. The development of oral functions such as mastication and speech may be accelerated if the patient grows without tooth missing as in this case.

To prepare the protecting plate, wax rims were made on the plaster cast at the height of the tooth crowns. An alginate impression was taken again on the plaster cast with the wax rims for the second cast, from which a plate was made with thermoplastic resin.

[Case 2]

In this case, the lower front teeth had been extracted for the purpose of preventing a bite wound. By 5 years of age, the wound had worsened by repeated tongue thrusting, the tongue surface was scarred and flat, and his taste sense probably had almost faded away. Habitual tongue thrusting to the space of the teeth missing is assumed as the background in this case. If a denture were applied to the space, such a severe condition could probably be avoided.

[Case 3]

A solitary lower front tooth produced an ulcer on the inferior surface of tongue which progressed and finally divided the tongue into two lobes. A denture was set on the lower jaw and the divided labes were sutured, after which ulceration ceased. This case also suggests that a well-arranged dental arch may prevent bite wounds to the tongue and other oral mucosa. However, we have experienced one case of bone exposure due to denture wearing. The prosthodontic treatment for the patient with this disorder is quite difficult, and requires close attention.

[Case 4]

A nine year old boy visited a dentist for left cheek swelling and fever. He was referred to a College Hospital for the treatment of mandibular osteomyelitis and cellulitis of mouth floor caused from the left lower first molar. Fig.37 is a radiograph taken at the Hospital. Antibiotics were immediately administered. The causal tooth was extracted a week later, a pathological jaw fracture occurred. The patient was referred to us at that time. At once, we performed an operation for

osteomyelitis, and the mandibular bone was fixed with wire. However, the boy removed the intermaillary wire anchorage (since he was mentally retarded and could not understand the need of fixation and endure the discomfort wiring in the mouth). A discrepancy was observed at the site of fracture again, and he had resumed to bite the lower lip probably, because of pressure to the mandibular nerve and residual numbness. After the osteomyelitis healed, an open reduction of the fracture was successfully performed with healing of the fracture. However, a screw of the plate on the mandibular edge came free and was loose in the subcutaneous tissue because the patient could not keep still. This case suggests how difficult fracture reduction can be for the patient with sensory neuropathy with anhidrosis.

Conclusion

A human being has five senses (hearing, vision, taste, smell, and touch) and receives these sensations through the ears, eyes, tongue, nose, and skin, respectively. These sensations are received, recognized, interpreted, and affect our learned and instinctive behaviour. A human being learns to control or modify his/her behaviour with the information received from these senses. Any of these senses may be disabled, and the inconvenience and hazards associated with the loss of vision or hearing are well known and have many therapeutic interventions. The sense loss of pain and temperature, although they may seem less significant, disables the recognition of innate and learned danger signals and thus also pose a threat to life itself.

The children who live in the world without the experiencing pain have difficulty learning new defense responses. Further their innate and instinctive responses to painful, dangerous stimuli is disabled. Consequently they suffer from repeated injuries and fractures and if they act without concern about their situation then these conditions will become worse.

To resolve these difficult problems, it is essential to diagnose the patient with sensory neuropathy with anhidrosis as early as possible to educate the family and the patient, to begin preventive efforts, and to start the early treatment of problems to avoid them becoming more serious. In these cases dentists have an important role together with pediatricians and orthopedists.

They say that medical service starts with knowing the patient's pain, while, for the patient with this disorder, it should start with understanding *the pain of lack of pain*.

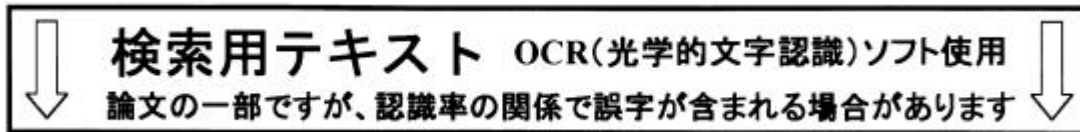
Self-inflicted bite wounds are the most serious dental problem for patients with this disorder. Many cases are difficult to treat. These may require the use of splints, tooth grinding, tongue suturing, and even tooth extraction. Regretably, there is actually no single dental treatment for self-inflicted bite wound in this disorder. As the patient may not be able to keep still because of hyperkineses or restlessness due to mental retardation, the dental treatment is difficult and prolonged in most cases.

Again, as there is no single therapy, all we can do is to conduct aggressive prevention and adequate and satisfactory symptomatic therapy. In practice, the tongue and cheek mucosa should be securely protected by early placement of a splint when a bite wound is found. In all cases, the guardian should be informed and instructed about self mutilation and the prevention of dental caries and periodontal disorders as soon after diagnosis as possible. Various interventions should be made to preserve morphology and function of the oral structure so that the patient can eat and taste more

normally.

Contributors

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研究要旨

先天性無痛無汗症の歯科的な問題として、舌や口唇、頬粘膜の咬傷、手指の咬傷、う蝕や歯周疾患に起因した上下顎骨髄炎、外傷による裂傷や顎骨骨折などが挙げられる。そこで咬傷および歯の早期喪失の予防のため保護プレートを着用し良好な結果を得ているが、長期にわたる保護プレートの使用で歯列、歯槽部の発育を抑制してしまう可能性があると考えられる。これについて生後 10 ヶ月から保護プレートを使用している女兒(症例 1)および乳幼児期に下顎前歯を抜歯された男児 2 例(症例 2,3)について歯列模型計測を行い検討した。

症例 1 では上下各歯牙間幅径は 4 才 9 ヶ月時、下顎乳犬歯部 $-1.1SD$ の他は著明に小さい部は認められなかった。症例 3 は永久歯で、上下顎共著明な前歯部の叢生が認められたが、下顎乳前歯 6 本早期に抜歯された既往があり、早期に多数歯を喪失することにより歯槽部の成長に影響した可能性があるかと推測された。