Oral Health Education in School for Children with Disabilities - Approach to Parents and Class Teachers -

Toshiko ADACHI[1*], Yoshiaki ONO[2], Akiko OHTANI1, Mikako KASAI[1], Osamu SHINOZUKA[3], Yuzo TAKAGI[2] and Zenko NAKAMURA[4]

[1]Section of Dental Hygiene, University Hospital, Faculty of Dentistry, Tokyo Medical and Dental Univ., Tokyo, Japan
[2]Developmental Oral Health Science, Graduate School, Tokyo Medical and Dental Univ., Tokyo, Japan
[3]Dentistry for Persons with Disabilities, Graduate School, Tokyo Medical and Dental Univ., Tokyo, Japan
[4]Metoropolitan Higashiyamato Medical Center for Disabled, Tokyo, Japan

Introduction

Children, parents and classroom teachers should understand that oral health and general health share an integral, mutual relationship. However, the oral health programs in most of the schools in Japan consist of annual dental examinations with only few oral health education curriculums. The oral health program at the school for children with disabilities attached to some university in Tokyo used to be conducted by a dentist and a nurse teacher. It contained only annual dental examinations for school children and making a notice of the results to the parents. Since the school year 1998, dental hygienists and nursing instructors have been involved in oral health education programs that may help the children, parents and teachers to understand the relationship between the mouth and the rest of the body.

Here, we are reporting the changes in the attitudes of the children, the parents and the teachers towards oral health activities after 2 year of this new program.

Materials and Method

The subjects studied were the children, the parents and the teachers in the kindergarten, elementary, junior high and high schools. The total number of the children in these schools was 70. Every child has some disabilities which come from mental retardation, Down's syndrome or autism. There were few children with movement disorder that would restrict their life styles, however, some children at the kindergarten and the elementary school needed assistance during their school lunch.

Information about the mental and physical condition of the children and behavioral features were obtained from the nurse teacher. The questionnaires included tooth brushing habits and the problems in tooth brushing or chewing of each child were given to the parents to answer in advance of the lectures each time (Table 1).

Table 1. Questionnaire before lecture

1. Who usually brushes the child's teeth?
2. When are the child's teeth brushed?
3. How much time is spent on child's brushing teeth?
4. Which method is used to help child's brushing teeth.
5. What kind of problems do you have about the mouth and teeth?

Information of each child about how to eat foods and problems in tooth brushing was acquired from the classroom teachers. Moreover, after two years of the program, a survey was conducted to determine how the parents understand the lectures and to know what the parents request more in the lecture in the kindergarten and elementary school (Table 2).

Table 2. Questionnaire after 2 years of the program
(This program started in 1998. Please answer the following questions.)

1. What was the most impressive topic of the past lectures for you?
2. What was the most interesting point about the lectures for you?
3. Style of lectures.
   Time distribution of the lecture on brushing the teeth: (too short / sufficient / too long)
   Total lecture time: (too short / sufficient / too long)
4. What theme would you like to be lectured on in the future?
5. What is your opinion of the past lectures and what are your requests for future lectures?

The school year in Japanese schools usually starts from April and ends in March next year. The designation of the year in this paper means the school year unless otherwise noted.

The present new program consisted of annual dental examination, individual instruction of tooth brushing after the lunch time, and lectures on the oral health to the children, parents and the teachers. The schedule of this program was shown in detail in Table 3 and Table 4. The curriculum of the program was customized according to the developmental stages of children and described in the following:

<Kindergarten>

The lectures on oral health were given to the parents of the kindergarten in 1998, 1999 and 2000. In each lecture, eating and speaking function and the facial expressions were explained. The anatomical

Table 3 Contents of Instructions (Kindergarten and Elementary School)

	Kindergarten	Elementary school
1998	February • Lecture to the parents Oral function and care - Oral function - Eating Oral health care -Keep the teeth healthy through the life - • Oral hygiene instruction - Check after brushing by a parent	Lower Classes in November • Lecture to the parents Swallowing and chewing - Oral functionrelated to eating - Training of oral function • Oral hygiene instruction - Check after brushing by a parent
1999	February • Lecture to the parents Oral function and care - Oral function related to eating - Oral health care • Oral hygiene instruction - Check after brushing by a parent	Upper classes in November • Lecture to the parents Swallowing and chewing - Oral functionrelated to eating - Training of oral function • Oral hygiene instruction - Check after brushing by a parent
2000	March • Lecture to the parents Oral function and care - Oral function (muscles aroud the mouth) - Oral health care • Oral hygiene instruction - Check after brushing by a parent	December • Lecture to the parents Swallowing and chewing - Oral function related to eating - Training of oral function • Oral hygiene instruction - Check after brushing by a parent - Order of brushing

features of the surrounding muscles of the mouth and their roles were also explained by using a windmill. The mouth cleaning exercise was performed for the children and parents after the school lunch.

< Elementary School >

The health educational instruction was given to the parents of the upper class children in 1998 and 1999. The main title of the lecture was 'Chewing' and the lecture was on the function of the mouth with the explanation of the relationship between the tongue and cheek movements during swallowing of the solid and liquid food. The mouth cleaning exercise was performed for the children and the parents after the school lunch in 1998, 1999 and 2000. For the purpose of improving the understanding of the children to master tooth brushing, graphic or poster presentation and instruction on how to move the tooth brush were performed for the children in the upper classes.

< Junior High School >

For the children in junior high school, the oral health education program was not possible in 1998 and 1999 because of the school schedule. The health educational instruction was given to the parents in 2000. The instruction contains lectures on some characteristics of the adolescent period, basic knowledge of caries and periodontal disease and their preventing measures. For the children, group instruction was performed using various educational materials such as pictures and drawings explaining

Table 4 Contents of Instructions (Junior high school and High school)

	Junior High Schhol	High School
1998		April • Lecture to the children Teeth and Oral disease - Dental caries and Periodontal Disease • Oral hygiene instruction to the children
1999		April • Lecture to the children Teeth and Oral disease - Dental caries and Periodontal Disease - Check of dental Plaque • Oral hygiene instruction to the children
2 000	December • Lecture to the children Teeth and Oral disease - Dental caries and Periodontal Disease • Lecture to the parents Oral Health and the features of adolescent • Oral hygiene instruction to the children	April • Lecture to the children Teeth and Oral disease - Dental caries and Periodontal Disease - Check of dental Plaque - Property of foods and dental plaque • Oral hygiene instruction to the children

the basics of caries and periodontal disease, and the preventing methodology. They are divided in small groups for the mouth cleaning exercise right after the lecture.

< High School >

The oral health instruction was only given to the children. The group instruction was performed after the annual examination of the teeth. The instruction contains the lecture on the basics of caries and periodontal disease, and preventing methodology. The visual aids such as pictures and posters were used to draw attention of children, since the motion of these aids focused the attention of the children. The colorful disclosing of the dental plaque with the pink disclosing solution was use for the children to help understanding how to brush properly in 1998. In 1999, the lecture contained on the knowledge how to check the dental plaque. In 2000, the lecture was on the relationship between the physical properties of food and dental plaque. Mouth cleaning instructions were given through the year 1998 to 2000.

Results

(1) Results of the questionnaires to parents before lecture

The results of the questionnaires to parents before lecture are demonstrated in the Table 5, Table 6, Table 7 and Table 8.

Table 5. Result of the questionnaire to kindergarten parents before lecture.

Year of survey		1998	1999	2000
Number of reply		5	4	7
Answer to Q1:	Parents	5	2	3
	Children	0	0	0
	Parents+children	0	2	4
Answer to Q2:	Morning	2	1	0
	Night time	0	0	0
	After meal	3	2	4
	Before sleep	3	2	3
Answer to Q3:	Less than 1 min.	1	0	0
	1 min. < 3 min.	1	1	3
	3 min. < 5 min.	1	2	2
	5 min. or more	3	1	1
Answer to Q4:	Lay down child	4	3	2
	In front of child	1	0	1
	No help	1	0	0
	Hands added	0	0	0

<Kindergarten >

Referring to Table 5, we received five replies from five parents in 1998, four of five in 1999 and 7 of seven parents in 2000. In Q1: Who brushes the child's teeth, five parents answered that they brushed the child's teeth in 1998, and three parents answered that they did, and four answered that they and the children did in 2000. In the Q2: When their children brush their teeth, the answers between 1998 and 2000 were not significantly different. For the Q3: How much time to spent for brushing, one brushed his teeth for less than one minute, one for over 1 minute but less than 3 minutes and two brushed for 5 minutes or more in 1998. During the year 2000, four brushed their teeth for over 1 but less than 3 minutes, one brushed for over 3 minutes but less than 5, and two brushed for 5 minutes or more. Three children in 1998 were not willing to have their teeth brushed, but only one child was uncooperative to parents brushing in 2000.

Table 6. Result of the questionnaire to lower classes parents before lecture.

Year of survey		1998	2000
Number of reply		6	9
Answer to Q1:	Parents	2	2
	Children	1	4
	Parents+children	3	2
Answer to Q2:	Morning	6	3
	Night time	0	0
	After meal	1	0
	Before sleep	2	8
Answer to Q3:	Less than 1 min.	1	1
	1 min. < 3 min.	1	3
	3 min. < 5 min.	2	3
	5 min. or more	2	1
Answer to Q4:	Lay down child	3	2
	In front of child	2	3
	No help	1	1
	Hands added	0	0

<Lower elementary school classes>

In the table 6, six out of six and nine out of 11 parents replied in 1998 and 2000, respectively. In the Question1, two parents answered that they did, one answered that the child did, and three parents answered that the both did in 1998. Two parents, four children by themselves, and three parents answered that the both did in 2000. For the Question 2, six children brushed their teeth in the morning, one after meals, and three before sleeping in 1998. Three children did in the morning, none after meals and nine children did before sleeping in 2000. It is noted that the number of children who brushed their teeth before sleeping increased. For the Question 3, in 1998, teeth brushing time was less than 1 minute (one), over 1 but less than 3 minutes (one), and over 3 but less than 5 minutes (two), and 5 minutes or more (two). In 2000, one brushed the teeth for less than 1 minute, four for over 1 minute but less than 3 minutes, three for over 3 minutes but less than 5 minutes, and one brushed for at least 5 minutes. Three children refused to allow their parents to brush their teeth in 1998, but only one child was uncooperative in 2000.

Table 7. Result of the questionnaire to upper classes parents before lecture.

Year of survey		1999	2000
Number of reply		9	7
Answer to Q1:	Parents	5	3
	Children	2	2
	Parents+children	3	2
Answer to Q2:	Morning	2	1
	Night time	0	2
	After meal	3	0
	Before sleep	7	4
Answer to Q3:	Less than 1 min.	0	0
	1 min. < 3 min.	5	4
	3 min. < 5 min.	2	3
	5 min. or more	3	0
Answer to Q4:	Lay down child	6	4
	In front of child	4	1
	No help	1	0
	Hands added	0	0

<Upper elementary school classes >

In the Table 7, nine of nine and seven of eight parents replied in 1999 and 2000. For the Question1, the answers were not significantly different between1999 and 2000. For the Question2, two children brushed their teeth in the morning, three after meals, none in the evening and seven before sleeping in 1998. One child brushed in the morning, none after meals, two in the evening and four before sleeping in 2000. For the Question 3, in 1999, no one brushed their teeth for less than 1 minute, five brushed for over 1 but less than 3 minutes, two brushed for 3 but less than 5 minutes, and two brushed for at least 5 minutes. In 2000, no one brushed teeth for less than 1 minute, four brushed for over 1 but less than 3 minutes, three brushed for over 3 but less than 5 minutes, and no one brushed for 5 minutes or more. In 1998, two children refused to allow their parents brush their teeth because the children disliked the process. No children had this problem in 2000.

<Junior high school >

According to Table 8, the answers in 2000 came from 19 of 21 parents. For Question 1, five parents brushed their child's teeth, ten parents answered their children did, and four parents said that they and their children did. For the Question 2, the children brushed their teeth as follows: 11 in the morning, 6 after meals, eight at night, and five before sleeping. For the Question3, two brushed their teeth for less than 1 minute, fourteen for over 1 but less than 3 minutes, and three brushed for over 3 but less than 5 minutes.

Table 8. Result of the questionnaire to junior high school parents before lecture.

Year of survey		2000
Number of reply		19
Answer to Q1:	Parents	5
	Children	10
	Parents+children	4
Answer to Q2:	Morning	11
	Night time	8
	After meal	6
	Before sleep	5
Answer to Q3:	Less than 1 min.	2
	1 min. < 3 min.	14
	3 min. < 5 min.	3
	5 min. or more	0
Answer to Q4:	Lay down child	9
	In front of child	1
	No help	0
	Hands added	3

(2) The results of the Questionnaire after two years of lectures

We administered the questionnaire after two years of lectures to the parents of the kindergarten and the elementary school in 2000. Fourteen of the 16 who attended the lecture responded to the questionnaire. For the Question 1: What they found was the most impressive topic of the lecture on oral health education, five parents answered oral hypersensitivity, five answered oral function, and three answered oral hygiene. For the Question 2: What contents they were interested in at the lecture, four parents answered movement of the oral muscles, two, using toothbrushes and two answered movement of the tongue. For Question 3 about the time spent for each topics in the lecture, two parents asked to lengthen the part regarding oral hygiene instruction, eleven thought that it was sufficient, and none wanted it shortened. For Question 4 about the length of the lecture, thirteen parents answered that they were satisfied. For Question 5 about what they thought about the lecture, the answers were that the explanation using visual aids was easy to understand, they could understand the points about brushing the teeth, and that receiving instruction on brushing the teeth was useful. For the question on what they wanted to hear in future lectures, five parents answered the relationship of the mouth to the rest of the body, three answered a dental clinic where they could receive a periodic dental examination, and one requested individual advice.

(3) Results of dental checkups for gingivitis

The results of the dental examination for gingivitis were as follows. Two children had gingivitis among fifteen in the elementary school, four of twenty-four in the junior high school and eight of twenty-eight in the high school in 1998. In 1999, one of seventeen children in the elementary school, twelve of twenty-one in the junior high school, and two of twenty-seven children in the high school had gingivitis. In 2000, two of eighteen children in the elementary school, twelve of twenty-one in the junior high school and four out of thirty children in the high school had gingivitis.

(4) Observations at school lunch

Throughout all classes, many children did not take a suitable amount of time to eat lunch. Some, who spent a short time eating lunch, did not masticate well and finished lunch within 5 minutes. Others, who spent a long time over lunch, could not close their mouths and could not swallow food well. Moreover, without the ability to eat the proper amount of food in one cycle of chewing, some put too much food into their mouths which made them unable to swallow it.

In the kindergarten and elementary school, in particular, many children could not use a spoon nor a fork well. Some could swallow food by lifting up their chin. Some leaked milk from their mouths and

could not swallow milk from a bottle. Some bent their back too much forward while eating and some leaked food from their mouths. Many children did not eat hard foods and refused to eat food that they disliked.

Discussions

In oral health education for the children with disabilities, the parents and the classroom teachers should have active participation to help and support the children to build up a healthy lifestyle; otherwise, it would be difficult for them to achieve this goal by giving instructions only at school. At the same time, proper instructions should be given to children according to their developmental stages.

Since there have not been many reports on the practical programs of oral health education in schools for children with disabilities, the brief description under this discussions, about this program approaching to the parents and teachers at this institution would be beneficial to those who deal with the similar type of problems despite the small number of the subjects for the present study. The number of the subjects should be increased in the future studies as well.

Before the implementation of oral health education program, we had received information from the parents, the classroom and nurse teacher and the theme and contents of lectures have bee selected according to the developmental stages of children.

(1) Instruction at the kindergarten and the elementary school

In a questionnaire to the parents before a lecture, they replied that children did not swallow food, or did not masticate food before swallowing it. Observations by the authors during school lunch revealed that some children did not masticate well, whereas some could not use their tongue well and could not mash food. Since these findings indicated that functional training with the knowledge of oral function was necessary, we raised the slogan for oral health education as "A Mouth is the Entrance to Health".

We lectured the parents and classroom teachers about the relationship between the mouth and the entire body and gave instruction about mouth cleaning in 1998 and 1999. Because the participating parents and teachers ate sesame, cookies and drank water at the lecture, they understood the relationships among the lips, tongue, cheeks and teeth, and among the mouth, eyes, and hands. Using similar experiments, they recognized that depending on the size and property of foods, different functions are required for mastication. They became interested in the function of the mouth by these experiences. When the importance of closing the lips and nasal breathing were discussed during the lecture that addressed the experience of swallowing water, one participant commented that their child often choked during meals. This problem was not discovered by the questionnaire, because the parents thought it was a common situation and not a problem.

We observed many children who could not swallow well during school lunch. Many children could not close their lips because they breathe through the mouth. We considered that this was associated with the fact that 11 of 21 children at the kindergarten and elementary school had rhinitis according to the medical examinations in 1999. We therefore taught them to blow the nose before meals and guided the parents to consult an otolaryngologist. Improvements were quite evident. The question concerning oral function from the classroom teachers noted this as a turning point.

In addition, during oral hygiene instruction, we saw many children who had rhinitis and who breathed through their mouths. We speculated that they could not breathe well through the nose and therefore refused to brush their teeth to avoid choking. We pointed out this problem to the parents and guided how to efficiently control brushing time. Consequently, many parents said that children stopped disliking brushing the teeth.

We made the parents recognize that oral function and oral hygiene have profound influences on the body. We considered that this was the starting point to think about the general health through oral health education and to approach from both school and home to improve the lifestyles of the children.

(2) Instruction at the junior high school

Instruction that considers the developmental stage of adolescence is needed for junior high school

children and their parents. However, we could not teach the children in 1998 and 1999 because of the school schedule. Twelve of 21 children (57%) had obvious gingivitis revealed by the dental examination in 2000. This was higher than that in the high (13%) and elementary (11%) schools, where the children had been given oral hygiene instruction.

Therefore we focused on improving the oral hygiene for these children. Although data from the nurse teacher indicated individual differences in the capabilities of gargling or brushing techniques, group instruction was performed for each class. Individual differences in ability were actually large during instruction. However, the children who could brush their teeth well gave advice to children who could not brush their teeth as well. Exchange of information between the children was surprisingly active. We would have to take this aspect into consideration in the future instruction session.

After reviewing the oral hygiene instruction to the children, we lectured the parents about the possible behavioral problems during adolescence, basic understanding of caries and periodontal disease, and how to prevent them. In the questionnaire before the lecture, 10 of 21 parents said that their children disliked their help and that the children were not aware of dental plaque. Some parents said that although they thought that the children's mouths did not become clean if the parents did not brush the children's teeth, the children refused their parents' help, so the parents tended stop participating in children's oral health care. We therefore advised the parents to make their children aware of plaque by using a dental plaque dyeing agent. Parents could point out which teeth were dirty without brushing the children's teeth, and ask the children to brush their teeth once again. Moreover, we also advised them to undergo a periodic dental examination with their family dentists, and gradually have the children manage their own oral health during adolescence.

(3) Instruction at the high school

We gave group instruction to high school children using the visual aids. The same medium was used in 1998 and 1999 from the viewpoint of the educational effect, and a new subject was added to that in 1999. Because we used the same visual aids, the children who saw then began to speak up about the contents of the lecture from the previous year. Thus, introducing instruction could become easier and the health education of the previous year could be effectively reviewed. Although 8 children among 28 had gingivitis in 1998, this decreased 4 among 30 by 2000. We did not lecture the parents of the high school children due to the conflicts with the school schedule. However, some parents participated in the lecture at the elementary school. We thought that in addition to a lecture, a method should be considered to pass on information about the mouth to parents.

(4) Dealing with oral health education at a special education school

To draw interest to the oral health care from the parents and their disabled children, a nurse teacher proposed that the 'health committee' of each classroom checked the toothbrushes every month to suggest when to change toothbrushes from 1998.

At the end of 1999, we presented the activity reports of 1998 and 1999, and the oral health educational curriculum according to school age (Tables 3 and 4). Tackling the issue as a school-wide activity, a staff meeting decided that "to promote the health of the mouth and eating function" is one of the requirements for the whole school. Because of this, not only the dentists, dental hygienists, and nurse teacher, but also classroom teachers, a dietitian, and a chef had participated in the activity. Consequently, "the school lunch should be chewed well" became one theme, and the teachers, a dietitian and chefs devised new school lunch menus in consideration of the functional status of the children.

Observations of children at school lunch showed that many children had incorrect posture while eating lunch and this led the children to spilling and cramming foods. The desks and chairs did not seem to suit the body of the children. We therefore asked the school to provide proper desks and chairs that is more suitable for the children. Some children used forks and spoons that were too large for them, which caused them to cram too much food into their mouths and for food to leak from their mouths. We therefore asked the children to bring spoons and forks from their homes. Finally, the children were equipped with utensils that suited them, such as a glass and a straw and all of these factors resulted in

the improvement of eating ability.

(5) Attitude Change at home

The ratios of children with gingivitis according to the results of dental examination in 1998 were 13%, 17% and 29% at the elementary, junior high and high school, respectively. However, these changed in 2000 to 11%, 57%, and 13%, respectively after teaching the classes. Gingivitis was not observed in the children at junior high school, who also attended instruction while in the upper classes of the elementary school. Moreover, many children brushed their teeth only by themselves or by the parents before instruction. However, after the instruction, the number of children and parents who brushed the teeth together increased (Table 5). Five children of the kindergarten and four of the elementary school, who had received instruction in 1998, also underwent more instruction in 2000. Although six of them brushed their teeth for less than 3 minutes in 1998, six of them did so for 3 minutes or more in 2000. This is considered that the parents' understanding of the mouth had changed owing to the instruction which resulted in the improvement of their children to do toothbrushing.

Although many children who brushed their teeth in the morning before instructions started in kindergarten and in elementary school, the number of children who brushed before sleeping increased in 2000. However, the number of children who brushed their teeth after meals did not increase, so we believe that instruction should be continued to bring further changes in oral hygiene.

In administering the new oral health programs at the school for children with disabilities, parental health education was found effective in the form of lectures, to improve the oral status of children and the following can be applicable to the program for the children without disabilities.

The parents' awareness of the problems and lecture information should be selected according to the answers of the questionnaires, rather than those determined by the usual lecture contents. Moreover, the satisfaction of the parents should be reached to make it easy for the children to accept the instruction. Thus, lifestyles might be improved in the short term if schools and homes share the same viewpoint about oral health education. School health education is an activity that aims to maintain and improve the health of children and school staff. Oral health education helps children to develop the capability and attitude to maintain healthy teeth and a healthy mouth. Therefore, practical activities that build up a view of total body health through the oral health education are required. We believe that we have come close to the ideal of school health through these activities.

Conclusions

We performed new oral health education program for three years at the school for children with disabilities attached to some university in Tokyo.

1) The teachers and the parents became interested and actively involved in oral health education.
2) The contents of instruction include not only caries prevention but also the oral function.

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Health Care of Adults with Prader-Willi Syndrome: A Questionnaire Study

Rika HIRAIWA[1,2]*, Akira OKA[1], and Kousaku OHNO[1]
[1] Division of Child Neurology, Faculty of Medicine, Tottori University, Yonago, Japan
[2] Eastern Shimane Rehabilitation Hospital for the Disabled, Matsue, Japan

Introduction

Prader-Willi syndrome (PWS), first described by Prader, Labhart, and Willi (1956), is a genetic disorder with the incidence of approximately 1 in 15,000 live births (Burd, L., et al. 1990 and Ehara, H., et al. 1995). It is caused by absence of expression of the normally active paternally-inherited genes within 15q11-q13, and its genotypes consist of paternal interstitial deletion (70-75%), maternal uniparental disomy (20-25%), and imprinting mutations (2-5%) (Ledbetter, D.H. et al. 1981, Butler, M.G. 1990, and Chotai, K.A. & Payne, S.J. 1998). The prominent features in PWS are infantile hypotonia, hypogonadism, short stature, mental retardation, behavioral problems, hyperphagia, and subsequent obesity (Holm, V.A., et al. 1993 and Cassidy, S.B. 1997). Behavioral problems and obesity intensify over time, contributing to serious health problems during adulthood. To investigate health issues, questionnaires were mailed to parents of PWS individuals in the cooperation with Japanese PWS society. Compiled the information of 29 individuals, who are18 years and older, we assessed their health problems and needs during adulthood.

Methods

Questionnaires were mailed to 369 parents of PWS individuals in the cooperation with Japanese PWS society. The questionnaires contained 40 structured questions with selection of choices and open space for comments, regarding (1) intelligence function; (2) living arrangements; (3) height and weight; (4) lifestyle: eating habits and physical activities; (5) medical conditions; (6) parents' views on health status and the needs for healthy life.

Results

Of the 369 mailed questionnaires, the information about 174 was received. A response rate was 47 per cent. Among 174 respondents, 29 were adults aged 18 or older. The 29 individuals with PWS, aged between 18 and 31 years, consisted of 14 males, 14 females, and one person whose gender was unanswered.

(1) Intelligence function

All, except for 4 unknown cases and 1 missing case, were retarded. The majority of subjects were moderately retarded, one third mildly retarded, and one was of borderline intelligence. (Table 1).

Table 1 Intelligence quotient

Intelligence quotient	*N*	*per cent*
Borderline (IQ 70-79)	1	3.4%
Mild (IQ 50-69)	8	27.6%
Moderate (IQ 35-49)	15	51.7%
Unknown	4	13.8%
No response	1	3.4%

(2) Living arrangements

Twenty-three PWS individuals (79.3%) lived at home. Four (13.8%) were institutionalized. A 21-year-old male had lived

in a commuter dormitory, but he was fired due to a troublesome social-relationship at work, and had to leave. Only a 25-year-old male resided at group home.

(3) Height and weight

The height and weight were not answered in one male. Among remaining 28 PWS adults, height ranged between 140-166 cm (mean, 152 cm) in the males, and between 131-147 cm (mean, 141 cm) in the females. The weight in the males and in the females ranged between 45-120 kg (mean, 77 kg) and 45-110 kg (mean, 68 kg), respectively. The mean body mass index (BMI) of the 28 adults was 33.5 kg /m^2 (range, 22.3-54.1 kg/m^2; normal range, 20-25 kg/m^2). 23 of 28 (82.1 %) were over-weighted with BMI more than 25 kg/m^2.

(4) Lifestyle

Eating habits

Majority of PWS individuals were thought to have a balanced diet; daily in 2 (6.9%), almost daily in 14 (48.5%), and occasionally in 3 (10.3%). However, a balanced diet was seldom taken in 2 (6.9%) and not often in 7 (24.1%). Most parents added personal comments expressing difficulty in dietary management. They tried various ways, such as locking on cupboards, not leaving food around, selecting low-fat/low-energy diet, preparing a meal with a plenty of vegetables to satisfy appetite, and so on. Parents were frustrated, in terms of protecting PWS individuals against overeating outside. The siblings as well as parents often felt stress to restrain themselves food when the PWS individual was around. Strict management of diet frequently made the mental condition of PWS persons unstable, aggravating eating problems like a vicious circle.

Physical activities

Table 2 showed habitual physical activities in our subjects. More than two thirds were in the habit of light to moderate physical activities such as taking a walk with a dog, running, swimming, and aerobic exercise. On the other hand, there were 5 (17.2%) people who hardly exercise. All of these 5 people were severely obese and the BMI of them ranged between 34.7 -54.1 kg/ m^2.

(5) Behavioral problems and psychiatric symptoms

Table 2 Habitual physical activities and the BMI

Physical activities	*N*	*per cent*	*BMI*
Often, moderate intensity	3	10.3%	22.3, 24.1, 34.5
Often, light intensity	16	55.2%	23.0-49.6
Weekly, light intensity	3	10.3%	26.3, 33.3, 35.6
Monthly, light intensity	1	3.4%	26.2
Rarely	5	17.2%	34.7-54.1
No response	1	3.4%	23.9

Table 3 Behavioral problems

Behavioral problems	*N*	*per cent*
Stubbornness	24	82.8%
Hyperphagia	24	82.8%
Food stealing	23	79.3%
Temper tantrums	22	75.9%
Lying	21	72.4%
Emotional labiality	19	65.5%
Self-injurious behavior (skin picking)	17	58.6%
Aggressiveness	17	58.6%
Repetitive speech	16	55.1%
Hypersomnia	15	51.7%
Laziness	10	34.5%
Stealing	7	24.1%
Compulsive behavior	6	20.7%
Passage related behavior disorder	6	20.7%
Wandering	5	17.2%
Pica	3	10.3%
Sexual behavior disorder	3	10.3%
Hyper activities	2	6.9%

Behavioral problems

Twenty-eight of 29 had some behavioral problems such as stubbornness, hyperphagia, food stealing, temper tantrums, lying, and emotional labiality (Table 3). Behavioral problems were not reported only in a 22-year-old female. She had received growth hormone therapy and was 147cm of height and 24.1 kg/m^2 of BMI. She had a balanced diet and enjoyed swimming once a week. Her parents thought she was in good health, though taking medicine for hyperlipidemia.

Table4 Psychiatric symptoms

Symptoms	*N*	*per cent*
Idleness, spiritlessness	6	20.7%
Delusion	6	20.7%
Depression	3	10.3%
Hallucination	3	10.3%
Manic psychosis	3	10.3%

Psychiatric symptoms

Some psychiatric symptoms during the past 5 years were reported in 11 of 29 (37.9%)(Table 4).

(6) Medical conditions

Medical check-up

All of 29 PWS individuals had their family physicians. Twenty-four (82.7%) have a check-up regularly; once per 1 or 2 months in 11, once or twice a year in 6, more than once a week in 3, a few times per month in 3, and no response about frequency in one. Other five people (17.2%) consulted doctors only when they had some complaints. While 10 (34.5%) could visit their familiar physicians by themselves, parents tended to accompany them for a check-up in 28 cases (96.5%) and institutional staff occasionally did so in 7 cases (24.1%).

Table 5 showed medical specialities that PWS individuals consulted in the past 5 years.

Table 5
The specialists whom peoples with PWS saw in the past 5 years

Specialists	*N*	*per cent*
Internist	19	65.5%
Dentist	19	65.5%
Pediatrician	18	62.1%
Dermatologist	15	51.7%
Ophthalmologist	15	51.7%
Otorhinolaryngologist	15	51.7%
Psychiatrist	10	34.5%
Orthopedic surgeon	9	31.0%
Surgeon	7	24.1%
Urologist	6	20.7%
Psychosomatic internist	2	6.9%
Rehabilitation	1	3.4%
Endocrinologist	1	3.4%

Diseases and symptoms

Diseases and symptoms in the past 5 years were shown in Table 6. Most PWS individuals were obese. The incidence of diabetes mellitus, fatty liver, hyperlipidemia, and sleep apnea among our patients in the past 5 years was 48%, 28%, 21%, and 7%, respectively. There were also high incidences of dermatological problems, such as impetigo and trichophytosis, otorhinolaringological diseases, such as otitis media and sinusitis, and dental problems like multiple caries. It seems to be related to the needs of dermatological, otorhinolaryngeal, and dental care (as shown in Table5).

Medication

Twenty-four (82.8%) took medicines for diseases associated with lifestyle, as well as behavioral and psychiatric disorders. The medicines were prescribed for diabetes mellitus in 10, psychic symptoms in 6, obesity (mazindol) in 4, behavioral problems in 3, hyperlipidemia, liver dysfunction, hyperuricemia, and constipation in 2, osteoporosis, epilepsy, urinary tract infection, and atopic dermatitis in one. Five diabetic patients required insulin therapy. One had tried sex hormone replacement and another (the 22-year-old female mentioned above) had received growth hormone treatment.

Hospitalization

Twenty-four of 29 (82.3%) had histories of hospitalization for 1-7 times in the past 10 years. The periods of hospitalization were from 1 week to 1 month in 11, from 3 to 6 days in 7, from 1 to 3 months in 5, within 2 days in 4, and more than 3 months in 2. Except 1 case with nephritis, the hospitalization for 1 month and more was aimed for weight control and treatment of diabetes mellitus.

(7) Views of family

Health status

While majority of parents thought that their child with PWS was healthy [6 (20.7%), very healthy and 10 (34.5%), healthy], about one third of people with PWS were considered unhealthy [not very healthy and mildly ill in 2 cases each (6.9%) and severely ill in 5 cases (17.2%)]. The 5 people considered severely ill, aged 18 to 25 years, were all obese with a BMI range from 33.3 to 35.6kg/m^2. However, there was no apparent relationship between health condition and the degree of overweight.

Problems on having a medical examination or hospitalization

Twenty-one parents replied about the problems on having a medical check-up or admission to a hospital (Table 7). One third of issues were related to behavioral problems of PWS individuals as well as poor understanding of the specific disorder by medical staffs.

Table 6

Diseases and symptoms in the past 5 years

Diseases and symptoms	*N*	*per cent*
Obesity	27	93.1%
Impetigo	15	51.7%
Richophytosis	15	51.7%
Diabetes mellitus	14	48.3%
Multiple carries	13	44.8%
Fatty liver, liver dysfunction	8	27.6%
Otits media	7	24.1%
Gingivitis	7	24.1%
Hyperlipidemia	6	20.7%
Sinusitis	6	20.7%
Constipation	4	13.8%
Hyperuricemia	3	10.3%
Bronchial asthma	3	10.3%
Hemorrhoids	3	10.3%
Osteoporosis	3	10.3%
Urinary tract infection	2	6.9%
Hypertension	2	6.9%
Sleep apnea	2	6.9%
Arthritis	2	6.9%

Table7 Problems at having medical check-up or admission to a hospital

Problems	*N*	*per cent*
Troubling others with behavioral problems	10	34.5%
No medical staffs who understand the specific disorder well	10	34.5%
Unsympathetic attitudes of others around	8	27.6%
Too long waiting time	5	17.2%
Expensive medical costs	4	13.8%
No appropriate medical facilities in the neighborhood	4	13.8%
Lack of cooperation on a check-up and examinations by a person with PWS	4	13.8%
Unavailableness of an accompanying person	3	10.3%
Insufficient equipments of hospitals and clinics	3	10.3%
No doctors who can continue to check-up	3	10.3%

The needs for healthy life

Twenty-eight of 29 parents checked some of the listed choices, in terms of the needs for healthy life (Table 8). Most parents also filled the blank with their voices. Some wanted the health care system for PWS patients without concerns of expensive medical cost. Majority of parents needed more information about PWS and also hoped that medical, educational, and social welfare professionals would deepen their understandings of the specific disorders. Some respondents desired mental care of PWS individual and family. One mother complained that she had to keep an eye to her son with PWS for 24 hours everyday, and it seemed to her harder than taking care of bed ridden, severely disabled people. Some parents expressed their concerns about how a child with PWS would live after they were getting older and gone. One mother wrote that there used to be little information about PWS when she was parenting and she wished young parents were provided with a better knowledge of PWS.

Table 8 The needs for healthy life

Needs	*N*	*per cent*
A guide book for people with PWS and their families	21	72.4%
Providing better knowledge about PWS to social community	18	62.1%
Hospitalization aimed for diet and exercise	17	58.6%
Medical coordinator	17	58.6%
Expanding of financial support for medical expenses	17	58.6%
Substantial day care service	16	55.2%
Regular medical check and a health card	13	44.8%
Support system for accompanying a patient	12	41.4%
Home visit system by medical staffs	3	10.3%

Discussion

Most of our subjects were short and over-weighted. The mean BMI was 33.5 kg /m^2 and 23 of 28 were with BMI more than 25 kg /m^2. In the study of 19 young adults with PWS (Höybye, C., et al. 2002), the mean BMI was 35.6 kg /m^2 and only three had a BMI less than 25 kg /m^2 despite a strict diet. Prevention of obesity in PWS is challenging because of the insatiable hunger and the reduced metabolic rate (van Mil, E.G., et al. 2000). In our survey, 14 of 29 (48%) were diagnosed as diabetes mellitus and 10 of the 14 diabetic patients were treated with drugs and 5 patients required insulin therapy. The prevalence of diabetes in our study was higher than previous reports. Greenswag, L.R (1987) reported among 232 adults with PWS aged between 16 and 64 years, 44 patients (19.0%) developed diabetes. In the other study of Butler, J.V. et al. (2002), 8 out of 32 (25%) PWS individuals, aged from 18 to 46 years, had diabetes. The difference of the incidence seemed due to the matter of sampling with different age distribution. The prevalence of liver dysfunction and hyperlipidemia were both above 20% in our subjects. About 80 % of people with PWS went to family physicians regularly and were taking medicine. In addition, they frequently need to receive psychiatric, dermatologic, otorhinolaryngeal, and dental care.

Whittington, J.E. (2001) reported that death rate in PWS population was about 3% per year. Significant morbidity might be induced by obesity and associated cardiovascular and pulmonary complications. The oldest person with PWS described in the medical literature is Miss AB who died aged 71 (Carpenter, P.K., 1994). She had been admitted for her last 16 years and her weight was

controlled at 71 kg with a BMI around 31 kg/m^2 by the staff diligence. This case emphasizes the needs for continued vigilance as to weight control throughout life.

It is important to note that only one person with PWS resided at group home and nearly 80 % of young adults lived at home, depending on their parents in our study. Most parents expressed difficulty in diet restriction and behavior management. The dual tasks of restricting food and managing behavioral problems might affect on family resources and energy. According to the survey of 232 cases with PWS (Greenswag, L.R, 1987) , the quality of family life markedly improved when the child with PWS left home.

There were a lot of voices of parents demanding better understandings of PWS in the community as well as promoting care of mental health of PWS individuals and their families. In addition to the education for the healthier life to PWS individuals and their family, it is essential for the community to share better knowledge about PWS (Greenswag, L.R. 1990).

Adults with PWS need self-supportive health services, providing diet management, exercise, and various leisure activities as well as medical examinations. Special facilities of the specific health care and group homes for PWS adults are recommended. In addition, appropriate management of diet from early childhood and approach for behavioral problems based on studies of cognitive and emotional characteristics underlying them, are requisite for health care of people with PWS.

Conclusion

Based on the information of 29 parents of adults with PWS on questionnaire, we investigated health issues and needs of PWS individuals during adulthood. The prevalence of diabetes mellitus, fatty liver , hyperlipidemia, and sleep apnea was 48%, 28%, 21%, and 7%, respectively. In addition, they frequently need to receive psychiatric, dermatologic, otorhinolaryngeal, and dental care. 28 of 29 exhibited some behavioral problems, such as stubbornness, hyperphagia, food stealing, temper tantrums, lying, and emotional labiality. 38% of subjects were reported some psychiatric symptoms. Obesity and behavioral problems were major concerns for the parents. Adults with PWS need the self-supportive health services, providing diet management, exercise, and various leisure activities as well as medical care.

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Fatal Outcome of a Severely Diabetic Patient with Prader-Willi Syndrome

Harumi SAIJO*, Takanori EZOE, Katsuhito ARAKI, Sui SONE, Hiroshi HAMAGUCHI, Harumi NAKAYAMA, Hisaharu SUZUKI, Yoshito HIRAYAMA and Masataka ARIMA
Tokyo Metropolitan Higashiyamato Medical Center for Severely Disabled, Tokyo, Japan

Introduction

Prader-Willi syndrome (PWS) is caused by the absence of the normally active paternally inherited genes in the q11-13 region of chromosome 15. Birth incidence of PWS is estimated at 1:15,000. PWS is characterized by infantile hypotonia, hypogonadism, short stature, characteristic facial features, cognitive delay, hyperphagia, increased risks of obesity, obsessive-compulsive and other behavioral problems (Table 1). Most patients with PWS show symptoms of hyperphagia and food-seeking behaviors. Complications associated with obesity still remain the leading causes of death in these patients even with early intervention. We report here with a patient of PWS, who died of uncontrolled diabetes mellitus (DM).

Case Report

The patient was a 24-year-old male with PWS. He was the third child of unconsanguineous healthy parent. He was born after 42-week gestation and his birth weight was 2970g. The muscle tone was low and he had difficulty in sucking milk. He could not control his head by the age of 6-month and could not walk alone until the age of 2. He was diagnosed as having PWS by the chromosomal analysis. Obesity became obvious at the age of 3. His parent started restriction of his food intake to prevent his obesity. However it resulted in loss of interest in playing and started to talk to himself all day. They gave up a diet for these adverse events. His body weight increased gradually and urine glucose became positive at the age of 13.

They visited our center for consultation when the patient was 15 years old. His height, weight and body mass index (BMI) were 147 cm, 76.5 kg and 35, respectively. Severe hyperglycemia was noticed. We recommended him to be hospitalized for a diet and treatment of DM. He was discharged after appropriate treatment. He visited our hospital again when he was 20 years old. His parent considered that they shouldn't force him to visit hospital because they thought that they should respect his decisions, although his cognitive level was that of 5 years old. We taught him the importance of a diet but it was difficult for him to control his appetite. We also educated his parent the importance of treatment. After graduation from the special school when he was 18, he started to work at a bakery shop. When he was 20 years old, cataract appeared and started to worsen. He suffered from recurrent cellulitis with severe edema and ulcer in his legs. Severe hyperglycemia persisted. His parent had given up to control his abandoned food intake. When he was 22, he was hospitalized several times for a treatment of cellulitis and a diet. He needed critical care for cardiac failure with pleural effusion. Hyperglycemia improved when he was treated in hospital. Unfortunately, his vision became poor due to cataract and diabetic retinopathy. Finally, his parent recognized the importance of the control of DM. The blood sugar level was well controlled with insulin therapy for three months after the discharge. We educated the importance of restricted access to food and close supervision on eating and spending money. However, it was quite difficult for his parent to change their life style. Hyperglycemia became out of control again and cellulitis relapsed at the age of 23. He was found dead when he was sleeping.

Discussion

Physical and psychological problems in patients with PWS are very severe. We would like to emphasize the importance to recognize various aspects of individuals with PWS and to support them.

Hyperphagia and obesity: Patients with PWS have difficulty in sucking and need tube feeding in infant. Obesity becomes obvious when they are three years old or so. Hyperphagia in PWS is considered to be due to an impaired satiety response. Recent data indicate anomalies in a specific set of oxytocin-secreting neurons in the paraventricular nucleus of the hypothalamus. It is thought that these neurons are related to satiety (Swaab, 1995). Treatment regimens consist of behavioral interventions such as low-calorie diets, exercise, restricted access to food and close supervision around food and spending money. We recommend diet in admission for two or three months. About 1200kcal-diet per day reduced the weight of individuals with PWS by approximately 10 kg during 3-months hospitalization.

Non-insulin dependent diabetes mellitus (NIDDM): PWS has been associated with morbid obesity and an increased propensity for early development of NIDDM. A reduced beta-cell response to glucose stimulation and a dissociation of obesity and insulin resistance are reported in non-diabetic PWS individuals in contrast to normal obese subjects (Schuster, 1996). Seventy-five % of Japanese adult patients with PWS suffered from NIDDM (Nagai, 1999). This percentage is higher than those of other countries (Butler, 2002). Oral agents and insulin therapy are not effective without restriction of food intake.

Ventilatory failure: Hypoventilation and desaturation during sleep are common, and sleep apnea may occur if obesity becomes marked. Hypercapnic respiratory failure is a common cause of death in PWS. These problems should be evaluated with sleep studies. Rapid weight loss must be achieved when hypoventilation is severe. It was reported nocturnal non-invasive ventilatory support improved nocturnal desaturation (Smith, 1998).

Leg ulceration and cellulitis: Skin picking is one of the self-injury behaviors in PWS. Lower nose, back pointer nail area, front legs are the specific injuring body locations (Symons, 1999) . We experienced some severely obese patients with PWS, who suffered from recurrent and diffuse cellulitis in their legs with ulceration. They were probably induced by frequent skin picking. Intravenous administration of antibiotics were necessary.

Cognitive function: The average IQ is approximately 65-70. It was reported that 27% of PWS patients showed moderate delays and 6% showed severe to profound delays. It was also reported that individuals with PWS scored better on visual motor discrimination skills than on auditory verbal processing skills (Curfs, 1991). It is well known that they often talk excessively and persevere verbally on a narrow range of topics

Maladaptive behavior: Behavioral problems associated with PWS are most noticeable. In addition to food-related difficulties, many maladaptive behaviors are seen such as tantrums, impulsivity, stubbornness, arguing with others, disobedience, stealing food or money to buy food, skin picking, compulsions, withdrawal and anxiety. Individuals with PWS show higher rates of compulsive symptoms such as hoarding, repetitive rituals, talking too much, and skin picking (Dykens, 1999). Several studies suggest that specific serotonin re-uptake inhibitors (SSRIs) are effective for some individuals with PWS to gain better control of compulsive symptoms (Dykens & Shah, 2003). Other problems are impulse control, psychotic and affective disorders. Depressive features such as sadness and low self-esteem, as well as anxiety and worries, are frequently noted in PWS. Treatment for

compulsivity and maladaptive behaviors consist of behavioral programming, a structured and predictable routine, family support and pharmacotherapy (Dykens & Shah, 2003).

We conjecture that the parent would be able to control their child in early childhood but it would become more difficult for them to cope with their child and to coordinate many medical and behavioral problems as they grew up. It was reported that a self-supporting program for children, youngsters and adults is effective (Descheemaeker, 1994). That program includes parental participation and education. We emphasize that a comprehensive and professional team approach is essential, especially to restrict caloric intake and provide psychosocial support for families.

In conclusion, we recognized the importance to support individuals with PWS and their family perseveringly.

Conclusion

1) We presented a patient with PWS, who had not received the appropriate treatment for obesity and DM.
2) The parent consulted the doctor in his early childhood but they didn't come to see the doctor regularly, because they respected his autonomy as he grew up.
3) We should support individuals with PWS and their family comprehensively with perseverance.

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