Oral and Dental Anomalies in Children with Down Syndrome at Queen Sirikit National Institute of Child Health

Parichart Sorathesn
Dentist (Expert level)
Dental Department
Queen Sirikit National Institute of Child Health
420/8 Rajvithi Road, Prayathai, Rajthavee,
Bangkok 10400
Tel: 02-3548325
Fax: 02-3548326
E-mail: ppom@health.moph.go.th

Abstract

The purpose of this study was to determine the prevalence and characteristics of oral and dental anomalies in children with Down syndrome. One hundred and four children (52 males, 52 females), who registered at Queen Sirikit National Institute of Child Health (QSNICH) were examined. The result showed that children with Down syndrome had stair palate, angle of the mouth pulled down, lower lip everted, lingual diastasis, scalloped and fissured tongue. In this study, the children had mouth closed and no tongue protrusion at rest, which resulted from manual stimulation on lips, cheek and tongue daily when they were infants. Tooth eruption was delayed in timing and sequence. For the majority of children, the first tooth erupted between age 9 to 16 months. The primary dentition was complete at the age 41 months or older. Missing teeth were found in 21.2% of the patients. Primary lateral lower incisors were the most frequently absent. Dental anomalies in shape such as peg shaped and geminated teeth were found at 4.8% and 2.8%. An irregular sequence of teeth eruption was common. In conclusion, Down syndrome children had abnormalities of palate, lips, tongue, an irregular sequence of teeth eruption, shape, size and number of teeth.

Key words: dental anomalies; Down syndrome; oral anomalies

Introduction

Down syndrome is a genetic disorder caused by abnormality at trisomy 21, which occurs 1 in 600 to 1000 births.[1] Previous reports showed several systemic manifestations.[2] The mortality rate of children with Down syndrome is 5-7 times higher than normal children and the life expectancy is 35 years.[3] The periods of the highest mortality risk are in infancy when congenital heart disease, leukemia and respiratory disease play a role. In adult, Alzheimer's disease and decline in immunological functions are found.[3]

Congenital cardiac anomalies is reported in 50% of infants with Down syndrome.[4] Death due to congenital heart disease was the highest during the first 2 years of life and only 40-60% live to age 10 years.[5] Gastrointestinal malformation occurred in 10-18% of patients.[6] Other findings included tracheoesophageal fistula, pyloric stenosis and duodenal atresia. Immunodeficiency in Down syndrome patients leads to an increased susceptibility to infection, and increased risk for developing neoplasia, particularly leukemia.[7]

Auto-antibodies against thyroid antigen were frequently found during early life. An increased prevalence of hypothyroidism has been reported.[8]

Allanson et al[9] studied the craniofacial pattern of Down syndrome patients which showed the midface was under developed, and the lips were broad, irregular, fissured, and dry.[10] An open mouth with protruding tongue was observed and the tongue appeared relatively large because of the small oral cavity.[11] Occasionally, true macroglossia may be present in addition to fissured tongue and geographic tongue has commonly been found.[12] The development of the midface was less complete than that of the mandible, resulting in reduction of the length, height and depth of the palate.[13] Significant reduction in length led to a stair palate appearance with high arch and occasionally palatal cleft like folds were found. The stair palate is often found in the first year of life in children with Down syndrome and it also occurs in premature infants and newborns with muscle hypotonia and sucking problem.[14] In normal children, it disappears from the second year of life, some children with Down syndrome showed a V-shaped palate.[15]

Eruption of both deciduous and permanent teeth was delayed in 75% of patients.[16] An irregular sequence of eruption was common.[17] Missing teeth have been reported at 23-47% of patients. Deciduous lateral incisors were absent at 12-17% of patients. Peg-shaped maxillary lateral incisors have been observed in 10% of patients.[18] Severe hypodontia (oligodontia) and anodontia have been noted occasionally.[19] Irregular alignment of teeth was common. Posterior crossbite, mandibular overjet, anterior openbite, crowded teeth and widely spaced teeth have been reported by several authors.[18,20]

Children with Down syndrome have morphological deviations that affect the dentition and the oral cavity. Most patients showed the typical open-mouth posture and protrusion of the tongue. In young patients, the open-mouth condition can be treated by Castillo-Morales orofacial therapy.[21,22] This therapy consisted of two parts: (1) manual stimulation and facilitation of lips, cheek and tongue (2) palatal stimulation with removable orthodontic plate modified by Castillo-Morales. Several studies in Germany, Sweden and Italy have been published on the positive effect of palatal plates in term of improving tongue position and lip activity.

In 2006, a multidisciplinary team for Down syndrome patients at Queen Sirikit National Institute of Child Health (QSNICH) was introduced, consisting of geneticist, surgeon, cardiologist, audiologist, ophthalmologist, orthopedologist, paediatric dentist, developmental medical specialist and physiotherapist. The team conducted the technology assessment for children with Down syndrome. In January 2007, the multidisciplinary team began the Down syndrome registry at QSNICH in order to collect data for studying and providing comprehensive care to these patients.

Since there was no previous study in Thailand, the purpose of this study was to determine the prevalence and characteristics of oral and dental anomalies in children with Down syndrome. The result of the present study will benefit to dental treatment plan for life long Down syndrome patients.

Materials and Methods

There were 178 children with Down syndrome who registered at QSNICH. The samples of this study were 104 children who had teeth erupted in oral cavities, aged 9 months to 62 months, 52 were females and 52 were males. All subjects were examined by one investigator. The procedure of oral examination was uniform, using mouth mirror and the patient seated in a dental chair. A tooth was considered to be clinically erupted when any position of its crown had pierced the gum. The data were recoded to facilitate statistical analysis by computer SPSS program version 16 for windows. Morphology of oral cavity was recorded in data form about lips, palate, tongue and dental anomalies. The parents were interviewed about behavior in terms of oral hygiene and oral stimulation. The protocol of this study was approved by the ethics committee of the QSNICH. After being informed of the objectives of the investigation, each parent provided written consent for the child to participate in this study.

Results

Stair palate appearance was apparently evident in the oral cavity of Down syndrome children. (Fig. 1A) The lips were fissured and dry. The angles of the mouth were pulled down with passive elevation of the hypotonic upper lip. The lower lip was also hypotonic and everted. (Fig. 1B) The tongue was also hypotonic. The midline junction of the tongue was weak (lingual diastasis) with excessive concavity of the frontal two-thirds of the tongue and weak frenulum. (Fig. 1C) Fissured and scalloped

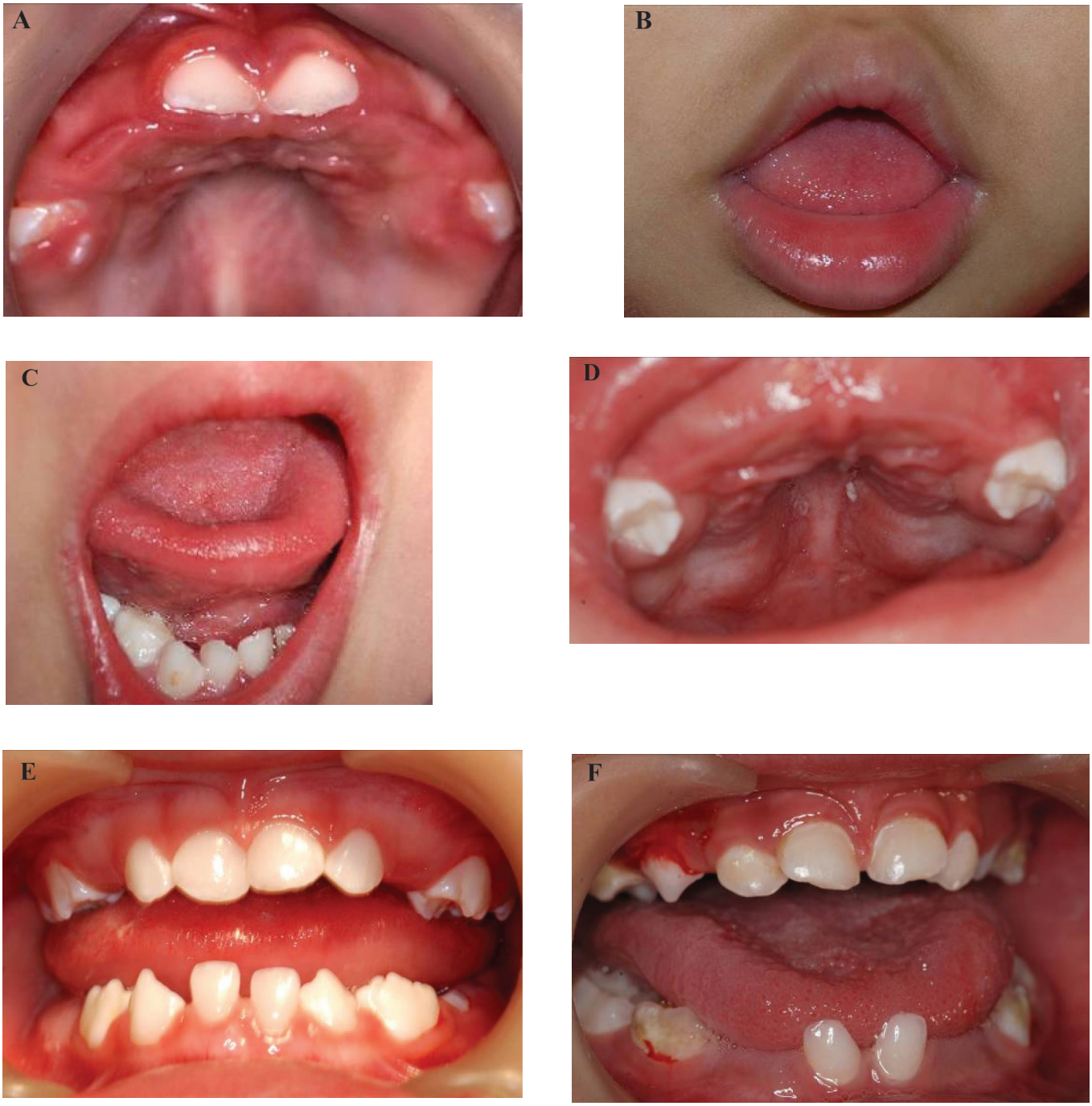

Fig. 1 *Oral and dental anomalies of children with Down syndrome*

Fig. 1A *22 month-old Down syndrome male with a typical "stair palate", which the prominent eminences have a step like transition to the palatal roof*

Fig. 1B *A19-month-old Down syndrome female with inactive upper lip and everted lower lip*

Fig. 1C *A 35-month-old Down syndrome male with a lingual diastasis that appears when the tongue is extended*

Fig. 1D *A 15-month-old Down syndrome female with deciduous first molar erupted first from the maxillary arch*

Fig. 1E *A 24-month-old Down syndrome female with missing deciduous lower lateral incisor*

Fig. 1F *A 26-month-old Down syndrome female with peg-shaped formation of deciduous lower central incisors*

Table 1 *Eruption of primary teeth in the children with Down syndrome*

Age (months)	Descriptive characteristics of erupted primary teeth amongs subjects (number of teeth)	
	Min - Max	Mean (s.d)
9-16	0 - 10	2.7 (2.7)
17-24	2 - 14	7.9 (3.5)
25-32	10 - 20	14.8 (3.6)
33-40	10 - 20	15.6 (3.2)
41+	17 - 20	19.4 (1.3)

Table 2 *Eruption of primary teeth in normal children (from Hatton).*[24]

Age (months)	Mean number of erupted primary teeth at various age ranges	
6	1	(33% have 1 teeth or more)
9	3	(80% have between 1 and 6 teeth)
12	6	(50% have between 4 and 8 teeth)
18	12	(85% have between 9 and 16 teeth)
24	16	(60% have between 15 and 18 teeth)
30	19	(70% have all primary teeth)

Table 3 *Irregular sequence of eruption of primary teeth of children with Down syndrome*

I	D		A		B		C			E
		D		A		B		C	E	
II	A	D			B		C			E
			A	D		B		C	E	

Table 4 *Usual sequence of eruption of primary teeth of normal children (from Meredith).*[26]

	A	B		D	C		E
A			B	D	C	E	

tongue was apparent in 28.8% (30/104) of children. The eruption period of primary dentition of the children with Down syndrome from this study was delayed in timing and sequence (Table 1) when compared with normal children from Hatton.[24] (Table 2)

Generally, the mean number of erupted primary teeth among the Down syndrome children in this study are lower than the normal eruption. That is the mean number of erupted teeth at age 9-16 months, 17-24 months, 25-32 months, 33-40 months and 41 months or above are 2.7, 7.9, 14.8, 15.6 and 19.4 respectively.

Regarding the sequence of eruption, an irregular eruption of the teeth of Down syndrome children has been found that the first decidous molar erupted first from the maxillary arch (Fig. 1D) in 2 cases and the first deciduous molar erupted after the deciduous central incisor in 3 cases. (Table 3) Comparing with the Meridith's sequence of normal eruption of primary teeth.[26] (Table 4) which the decidous lower central incisor is normally erupted first.

Regarding the dental anomalies, missing teeth of primary dentition were found in 21.2% (22/104) of the patients compared to the eruption period of normal children. The missing teeth were lower deciduous lateral incisors (Fig. 1E), lower deciduous central incisors, upper deciduous lateral incisors and lower deciduous canine, in descending order (Table 5). Geminated teeth were found in 2.8% (3/104) on lower lateral incisor and canine, lower central incisor and lateral incisor of the primary teeth. Peg-shaped formation was found in 4.8% (5/104) on upper lateral incisor and lower central incisor of the primary teeth. (Fig. 1F)

Table 5 *Missing teeth of children with Down syndrome*

Missing teeth	Cases (n)	Percentage
#72, #82	10	45.5
#72	4	18.2
#71, #81	2	9.1
#71, #72, #82	2	9.1
#52, #62, #72, #82	2	9.1
#71	1	4.5
#83	1	4.5
Total	22	100

Discussion

Previous studies reported the prevalence of missing teeth in the children with Down syndrome were 12.7 to 50%.[2,27] Agenesis was 10 times more than normal population.[19] The eruption period was delayed with irregular sequence. The first eruption was usually at the age 12 to 14 months but can be delayed to age 24 months. Eruption of primary teeth may not

be completed until age 48-60 months[28], compared with 30 months in healthy children. This study showed missing teeth at 21.2%. The first eruption was 9-16 months and complete eruption was 41 months and up. The children with Down syndrome had stair, v-shaped, high vault and short palate which reduced the retention of maxillary denture. Orthodontic and surgical correction, prosthodontic appliances or implants should be considered in older patients. The patients had fissured tongue which retained food and caused halitosis. Thus,brushing of the dorsal surface of the tongue should be recommended. However, the children with Down syndrome at QSNICH had mouth closed and no tongue protrusion at rest, which resulted from manual stimulation for lips cheek and tongue without using palatal plate. Fischer-Brandies[21] and Limbrock et al[22] reported that the use of palatal stimulation plates appear to result in a decrease in tongue protrusion in successful cases, but have little effect on lip posture or mouth closure.[29] Whilst inagreement that palatal plate therapy had no effect on hypotonia[30], Backman et al[31] found that such appliances when inserted at the age of 6 to 18 months may be beneficial to oral motor performance and in preparation for the development of speech. Since the children with Down syndrome had delay teeth eruption and had less efficient chewing, their parents should consider for proper food for them. Down syndrome children normally have mouth breathing and respiratory tract infection, causing them to have severe chronic periodontitis, which possibly occur with below or 6-year-old Down syndrome children.[32]

It is important for parents of children with Down syndrome to be instructed on proper oral care early in life. The application of topical and systemic fluoride and occlusal fissure sealants are recommended. Carious lesions in the primary dentition should be properly treated. It is also critical to maintain the primary dentition as long as possible. As for the dentist, dental treatment for children with Down syndrome should be considered that the incidence of upper airway obstruction is as high as 31%.[33] Down syndrome patients show increased laxity of transverse ligaments between the atlas and the odontoid process of the cervical vetebrae and between the altas and occipital condyles at the base of the skull.[30] Injury can occur with hyperextension or radial flexion of the neck or direct pressure on the neck or upper spine, leading to irreversible spinal cord damage. Thus, dentists should be aware of the risk during manipulation of the patient's neck when there is a positive history of atlantoaxial laxity. Furthermore, dentist should consider for infective endocarditis and heart failure due to high incidence rate of congenital cardiac defect up to 50%.[5] Multidisciplinary team should detect the syndrome early and plan to take care of those patients for long term life with well-being.

Conclusions

In this study, the children with Down syndrome had several oral anomalies including stair palate, angle of the mouth pulled down, lower lip everted, lingual diastasis, scalloped and fissured tongue. Dental anomalies consisted of delay in timing and sequence of teeth eruption, missing teeth, peg-shaped teeth and germination.

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บทวิทยาการ

ลักษณะความผิดปกติของอวัยวะในช่องปากและฟันในเด็กกลุ่มอาการดาวน์ ในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี

ปาริชาติ สรเทศน์
ทันตแพทย์เชี่ยวชาญ กลุ่มงานทันตกรรม
สถาบันสุขภาพเด็กแห่งชาติมหาราชินี
420/8 ถนนราชวิถี แขวงทุ่งพญาไท เขตราชเทวี
กรุงเทพฯ 10400
โทรศัพท์: 02-354-8325
โทรสาร: 02-354-8326
อีเมล: ppom@health.moph.go.th

บทคัดย่อ

การศึกษาครั้งนี้มีวัตถุประสงค์ เพื่อหาอัตราการเกิดความผิดปกติ และลักษณะความผิดปกติของอวัยวะในช่องปาก และฟันในเด็กกลุ่มอาการดาวน์ที่ได้ลงทะเบียนเป็นคนไข้ในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี ทันตแพทย์ได้ตรวจเด็กกลุ่มอาการดาวน์ที่เริ่มมีฟันขึ้นในช่องปากจำนวน 104 คน เป็นชาย 52 คน และหญิง 52 คน พบว่าภายในช่องปากของเด็กกลุ่มอาการดาวน์จะมีเพดานปากขนาดสั้นกว่าเด็กปกติ และมีลักษณะรูปร่างเป็นขั้นบันได ริมฝีปากมีลักษณะเป็นร่องแตกแห้ง มุมปากอักเสบ ลิ้นมีอาการอ่อนแรง มีลักษณะเป็นร่อง และมีรอยหยักด้านข้างลิ้น เด็กในกลุ่มที่ศึกษานี้ ได้รับการนวดกระตุ้นริมฝีปาก แก้ม และลิ้น ตั้งแต่ยังเป็นทารก ทำให้หุบปากได้สนิท ลิ้นไม่ยื่นออกนอกปาก การขึ้นของฟันจะช้ากว่าเด็กปกติ ฟันซี่แรกเริ่มขึ้นเมื่ออายุ 9-16 เดือน ฟันขึ้นครบ 20 ซี่ เมื่ออายุประมาณ 41 เดือนขึ้นไป ในเด็กกลุ่มอาการดาวน์ร้อยละ 21.2 มีฟันที่ขาดหายไป ฟันที่หายไปที่พบมากที่สุดคือ ฟัน #72, #82 (ฟันตัดซี่ข้างล่างซ้ายและขวา) พบฟันมีรูปร่างผิดปกติ เช่น มีลักษณะแหลมเล็ก ร้อยละ 4.8 และพบฟันแฝด ร้อยละ 2.8 ในเด็กกลุ่มนี้มักพบว่าลำดับการขึ้นของฟันแตกต่างไปจากเด็กปกติ โดยสรุปพบว่าเด็กกลุ่มอาการดาวน์กลุ่มนี้มีความผิดปกติของเพดานปาก ริมฝีปาก ลิ้น ความผิดปกติเกี่ยวกับการขึ้น รูปร่างของฟัน และจำนวนฟัน