Effects of Dysphagia Treatment Applied to Infants with Pierre Robin Syndrome

– Single Subject Research Design

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Abstract

Pierre Robin syndrome is characterized by micrognathia, glossoptosis, and cleft palate. Infants with Pierre Robin syndrome cause feeding difficulty, upper airway obstruction, and other symptoms. This study aims to examine the effects of applying dysphagia treatment to infants with Pierre Robin syndrome. The study participant was an infant who was born four weeks premature and referred for dysphagia treatment approximately 100 days after birth. At the initial assessment, the infant showed oral sensory sensitivity, a high level of facial and masticatory muscle tension, and a low stability of the chin and cheeks with almost no normal “sucking-swallowing-breathing” pattern. We set the baseline period and intervention period using the AB design. During the baseline period, non-nutritive sucking training using a rubber nipple was conducted without implementing an oral stimulation intervention. During the intervention period, non-nutritive sucking training and an oral stimulation intervention were performed. After the intervention period, the infant’s daily oral intake and oral intake per time significantly increased compared to that during the baseline period. We observed that the oral intake time of the infant decreased during the intervention period compared to that in the baseline period, which indicated an improvement in control over the chin, tongue, and lip movements, a change in muscular tension, and stabilization of the “sucking-swallowing-breathing” pattern. We provided dysphagia treatment before breastfeeding, it was positive effects such as normal development of the infant, transition from tube feeding to bottle feeding, and enhancement of overall oral motor function.

Keywords: Pierre Robin Syndrome, Infant, Dysphagia, Oral stimulation intervention,

1. INTRODUCTION

Pierre Robin syndrome is characterised by micrognathia, glossoptosis, and cleft palate. The cause of this disease has not been clearly identified, but it has been reported that the disease has an incidence between 1:2000 and 1:5000, and it is assumed to have an autosomal recessive inheritance pattern [1]. In 1911, Shukowsky observed a combination of the above three symptoms in a newborn baby that caused dyspnoea and
cyanosis, which posed a grave threat to the infant’s life. The disease was identified and named by Pierre Robin in 1923 [2]. Infants with Pierre Robin syndrome have a very small lower chin, shortened tongue muscles with the tongue pulled back and down, and an incompletely sealed palate that causes feeding difficulty, upper airway obstruction, and other symptoms. Infants can also experience complications such as intra-airway aspiration with resulting lung infections [3]. In addition, cardiovascular disorders such as pulmonary stenosis, patent ductus arteriosus and atrial septal defect, lung dysfunction, musculoskeletal deformity, malformation of the fingers and toes such as polydactyly and oligodactyly, and exotropia are occasionally observed. In many cases, the syndrome also includes epilepsy, problems with muscular tension, and speech disorders due to problems in the central nervous system [4].

Many newborn infants with Pierre Robin syndrome experience dysphagia. The normal swallowing process consists of an oral phase, pharyngeal phase, and esophageal stage. Dysphagia collectively refers to the problems that occur during the movement of food from the mouth to the stomach [5]. Most patients with dysphagia show dysfunctions in the oral phase and pharyngeal phase that lead to residue and aspiration in the esophageal stage. This causes various complications, including malnutrition, aspiration pneumonia, and dehydration, and in the most serious cases, death [6]. As such, dysphagia can deter the growth of a child and, more importantly, can put a child’s life at risk. Early intervention of any treatment is very important [7]. Especially for infants, eating is closely related to quality of life as well as survival. Intervention for eating is also important for the future development of the child's emotional growth [8].

Oral and pharyngeal muscles need to contract and expand harmoniously during the normal swallowing process. However, infants with Pierre Robin syndrome have chronic oral-function issues due to micrognathia, glossoptosis, and cleft palate. Many infants cannot complete the normal swallowing process since their nerves are impaired. As a result, they have abnormalities in the tension of the lips, tongue, and masticatory muscle and abnormal oral motor skills. An operation to secure the airway can be conducted on a newborn baby. However, the corrective operation for cleft palate is generally conducted at approximately 10 weeks after birth followed by staphyloplasty at 12 to 18 months after birth. As a result, infants can still feel uncomfortable with their oral structure when eating meals until the staphyloplasty is completed [9]. In particular, instability in the chin and cheeks, and abnormalities in the tension of the lips, tongue, and masticatory muscle can exacerbate the oral structure challenges, so consistent intervention therapy must be simultaneously offered.

If an infant displays a problem with oral feeding, nasal-gastric tube feeding (NG tube feeding) is usually started. NG tube feeding is a method used to improve the nutritional status of malnourished patients and to help effectively provide nutritional support. However, as the nutritional intake period using a NG tube feeding becomes longer, an infant can become susceptible to adverse effects, including aspiration or abnormal oral reflexes [10]. Therefore, it is important to transition infants to self-feeding by training them to suck and swallow independently. Studies have been consistently conducted on non-nutritive sucking and oral stimulation intervention to enhance oral feeding of infants with weakened swallowing function; in particular, oral stimulation intervention has been presented as an efficient way to alleviate dysphagia [11].

Pierre Robin syndrome is a rare disease; therefore, there are not many clinical cases. Although there are several studies on surgeries to secure airways and the treatment of dental caries, it is difficult to find studies on the treatment of infant dysphagia. Most infants with dysphagia experience malnutrition and growth retardation due to their swallowing problems; therefore, it is very important to treat dysphagia. In this study, a paediatric occupational therapist carried out oral stimulation intervention in an infant with Pierre Robin syndrome to treat their dysphagia, and this study aims to examine the results and how intervention therapy helped the infant.

2. METHODS

2.1. Participants

2.1.1. Past Medical History
The infant in this study was a girl born on March 3, 2017. The mother of the infant was hospitalized twice
in the M obstetrics and gynaecology clinic in January owing to preterm labor, and she was treated with antibiotics from February 27 to March 2 because of amniotic fluid leakage. Thereafter, she visited the clinic on March 3 for a check-up and was diagnosed with hypertension. Therefore, an emergency caesarean operation was conducted, and she gave birth to a baby at 35 + 5 weeks’ gestation (Bwt. 2140 g). At the time of birth, the baby was A/S 9/1, but the initial oxygen saturation was 85%. After applying 10 L O2, the oxygen saturation was maintained at 100%. However, the baby was transferred to the NICU of S hospital due to respiratory distress, since the baby showed chest retraction and nasal flaring. The doctor observed cleft palate, micrognathia, and glossoptosis at the time of transfer, so the infant was diagnosed with Pierre Robin syndrome.

Figure 1 shows the oral structure of a child with cleft palate. Tube feeding was initiated at the NICU. Desaturation occurred several times, and respiratory treatment was conducted accordingly. As the infant suffered from airway oedema, the airway was secured by temporary intubation. The infant stabilized after an operation to secure the airway on May 18, 2017, and a request for swallowing treatment was made on June 23, 2017 from the department of rehabilitation medicine.

2.1.2. Swallowing Function

The swallowing function of the infant was assessed at 100 days after birth. Several assessment tools are used to observe infant feeding, but most of the tools lack standardization. In addition, these assessment tools are usually applied when the patient is age 1 year or more [12]. Therefore, we conducted our assessment using basic observation. NG tube feeding was being provided to the infant at the initial assessment stage. In the oral-motor function assessment, the oral sensory sensitivity and the tensions of the facial muscle, orbicularis oris muscle, and masticatory muscle were recorded as high and the stability of the chin and cheeks was recorded as low. The position of the tongue was observed to be pulled backward and muscle tone was high. “Sucking-swallowing-breathing” patterns were hardly observed, and the swallow reflex was very weak. The infant took in nutrition by 80 cc per time at a total of 8 times per day, entirely by NG tube feeding.

2.2. Research Environment and Design

2.2.1. Research Environment

In this study, the dysphagia treatment was conducted in the infant’s incubator at the neonatal intensive care unit (NICU) of S hospital. During the baseline period, only non-nutritive sucking training was performed using a rubber nipple, and attempts were made to feed the infant using an ordinary feeding bottle. If oral feeding was not possible with an ordinary feeding bottle, NG tube feeding was implemented to provide supplemental nutrition. During the intervention period, non-nutritive sucking training and oral stimulation intervention were synchronously conducted, and a special feeding bottle was used. Similar to that in the baseline period, NG tube feeding was implemented if oral feeding was not possible.

2.2.2. Research Design

This study is a single-subject research design using the AB design. The research period was from March 15 to April 7,
2017, and measurements were conducted for a total of 25 sessions. During the baseline period, non-nutritive sucking training was performed, and the infant took in nutrition through oral feeding using either an ordinary feeding bottle by a nurse or NG tube feeding. During the intervention period, non-nutritive sucking training and 30-minute oral stimulation intervention was conducted; thereafter, oral feeding using a special bottle or NG tube feeding was offered similar to that in the baseline period.

2.3. Instruments

2.3.1. Assessment of Feeding Volume
To assess the feeding volume, this study used an ordinary infant feeding bottle (Figure 2) and a special feeding bottle (Figure 3). During the baseline period, we assessed the feeding volume using a Green Mom PP Feeding Bottle for newborn babies with a total volume of 140 mL. During the intervention period, we used a Medela Special Feeding Bottle manufactured by Medela AG for infants with cleft lip and palate and premature babies with a total volume of 150 mL. The special feeding bottle is designed with a long nipple so that infants with a cleft lip and palate can easily suck the contents. Each bottle has a minimum measuring unit of 10 mL.

![Figure 2. Ordinary nipple of feeding bottle](image2)

![Figure 3. Special nipple of feeding bottle](image3)

2.4. Research Process

2.4.1. Baseline Period
During the baseline period, non-nutritive sucking training was offered, and oral stimulation intervention was not performed. Feeding was attempted every four hours with an ordinary feeding bottle. Any residual content was given by NG tube feeding. We measured the feeding volume the infant took orally from the feeding bottle and, to minimize errors in measurement, we did not provide any treatment stimulation while measuring the feeding volume. Feeding was conducted in the incubator following the feeding schedule to minimize any environmental factors that could influence the infant’s swallowing.

2.4.2. Intervention Period
During the intervention period, we conducted non-nutritive sucking training and 30-minute oral stimulation intervention, and then fed the infant using a special feeding bottle. Oral stimulation intervention was performed five times per week for three weeks for a total of 15 times. Oral stimulation intervention could not be offered at the 11th, 12th, 18th, 19th, and 25th sessions due to hospital holidays, so feeding was provided by using a special feeding bottle after conducting non-nutritive sucking training. For those five sessions, therapy intervention was not offered, but the data for the sessions has been included as it can be used to prove the effects of dysphagia treatment when only a special feeding bottle is used without conducting oral stimulation intervention.

2.4.3. Intervention Details
The dysphagia treatment program was customized for the infant in this study by referring to the “Oral stimulation program” of Sandra [13], which implemented treatment for preterm infants receiving tube feeding
to help them transition to oral feeding (Table 1). The program consisted of methods to stimulate the neck, chin, cheeks, upper lip, lower lip, upper gum, lower gum, internal cheeks, and tongue-surrounding parts for 30 minutes at a time. After conducting the treatment program, feeding was offered by using the Medela special feeding bottle at the scheduled time. In this study, we used a special feeding bottle during the intervention period to determine whether the unique and longer nipple design is helpful for infants with cleft palate issues. Treatment was provided by an occupational therapist with more than 10 years’ experience in paediatric dysphagia treatment and education, and the difficulty level of activities was adjusted depending on the condition of the infant.

Table 1. Oral stimulation program

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<th>Structure</th>
<th>Stimulation steps and purpose</th>
<th>Frequency /duration</th>
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| Neck               | • Inflect and relax the excessive neck extensor  
→ Maintain neck at the centre position and extend hamstring | N/A /3 min          |
| Chin               | • Slightly inflect the head to keep the centre position arrangement and make the stable   
→ Secure the stability of the chin | N/A /2 min          |
| Cheek              | 1. Place index finger at the base of the nose. 
2. Compress the tissue, move finger toward the ear, then down and toward the corner of the lip (ex, C pattern). 
3. Repeat for other side.  
→ Improve range of motion and strength of cheeks, and improve lip seal | 4× each cheek /2 min |
| Upper lip          | 1. Place index finger at the corner of the upper lip. 
2. Compress the tissue. 
3. Move the finger away in a circular motion, from the corner toward the centre and to the other corner. 
4. Reverse direction.  
→ Improve lip range of motion and seal | 4× /2 min           |
| Lower lip          | 1. Place index finger at the corner of lower lip. 
2. Compress the tissue. 
3. Move the finger away in a circular motion, from the corner toward the centre and to the other corner. 
4. Reverse direction.  
→ Improve lip range of motion and seal | 4× /2 min           |
| Upper and lower lip curl | 1. Place index finger at centre of lip. 
2. Apply sustained pressure, stretch downward toward the midline. 
3. Repeat for lower lip-apply sustained pressure, and stretch upward toward the midline  
→ Improve lip strength, range of motion, and seal | 2× each cheek /2 min |
| Upper gum          | 1. Place finger at the centre of the gum, with firm sustained pressure slowly move toward the  
→ Improve range of motion of tongue, stimulate swallow, and improve suck | 2× /2 min           |
2.5. Analysis Methods

The collected data were analysed using Microsoft Office Excel 2007 and SPSS ver. 20.0. The feeding volume was checked every day during both the baseline period and the intervention period, and the frequency analysis was conducted using the daily check results. The measurements obtained during the baseline period and the intervention period were compared and analysed using visual graphs and descriptive statistics.

3. RESULTS

3.1. Change of Average Oral Feeding Volume per Time

The average oral feeding volume per time is a value calculated by dividing the total daily oral feeding volume by the number of feeding times and visually presented it (Figure 4). The average oral feeding volume per time during the baseline period was 1.6 ± 0.4 mL, and the average oral feeding volume per time during the intervention period was 17.6 ± 7.2 mL. The average volume increased 16.0 mL during the intervention period compared to the baseline period. The average value of the 11th, 12th, 18th, 19th, and 25th sessions when only a special feeding bottle was used was 13.5 ± 6.1 mL, which was an 11.9 mL increase from that in the baseline period.
period average but approximately 4.1 mL less than the intervention period average.
During the intervention period, all 20 data sources were above the mean + 2 standard deviations of the baseline indicating that dysphagia treatment positively affected the change in oral feeding volume. The maximum value, minimum value, and median value of the total oral feeding volume on the box plot were distributed in the region with the larger oral feeding volume when the dysphagia treatment and special feeding bottle were applied together compared to when only the special feeding bottle was used (Figure 5).

3.2. Change of Total Daily Oral Feeding Volume
We measured the change in total daily oral feeding volume and visually presented it (Figure 6). The average oral feeding volume during the baseline period was 4.8 ± 1.1 mL, and the average oral feeding volume during the intervention period was 35.3 ± 2.0 mL. The average volume increased by 30.5 mL during the intervention period compared to the baseline period. The average volume at the 11th, 12th, 18th, 19th, and 25th sessions when only a special feeding bottle was used was 25.0 ± 17.6 mL, which was a 20.2 mL increase from the baseline period average but approximately 10.3 mL less than the intervention period average, when the dysphagia
treatment was performed. During the intervention period, all 20 data sources were above the mean + 2 standard deviations of the baseline, indicating that the dysphagia treatment positively affected the change in oral feeding volume. The maximum value and median value of the total oral feeding volume on the box plot were all distributed in the region with the larger oral feeding volume when the dysphagia treatment and special feeding bottle were applied together compared to when only the special feeding bottle was used (Figure 7).

![Figure 6. Change of total daily oral feeding volume per session](image)

![Figure 7. Change of total daily oral feeding volume compared by intervention method](image)

3.3. Feeding Time for 1 cc Volume
We measured the feeding time required to take in a volume of 1 cc and visually presented it (Figure 8). The average feeding time during the baseline period was 1.7 ± 0.3 minutes, and the average feeding time during the intervention period was 1.1 ± 0.1 minutes. The average feeding time decreased by 0.6 minutes during the intervention period compared to the baseline period. The average feeding time at the 11th, 12th, 18th, 19th, and 25th sessions when only a special feeding bottle was used was 1.2 ± 0.1 minutes, which was an approximate 0.5-minute decrease from the baseline period average.

During the intervention period, all 20 data sources were above the mean + 2 standard deviations of the
baseline, indicating that dysphagia treatment reduced the infant’s feeding time. There was no significant difference in feeding time when the dysphagia treatment and special feeding bottle were applied together compared to when only the special feeding bottle was used (Figure 9).

4. DISCUSSION

Pierre Robin syndrome is a congenital disease characterised by micrognathia, glossoptosis, cleft palate, and respiratory obstruction [14]. Rather than hereditary, this disease is characterized by serial reactions, wherein micrognathia causes glossoptosis followed by upper airway obstruction; therefore, it is also referred to as Pierre Robin sequence [15]. Patients with this syndrome have chronic oral function problems because of structural deformities of the oral cavity, and many newborns experience difficulties in the normal swallowing process. As a result, dysphagia is a life-threatening issue, and the disease should be promptly treated [9].

The infant in this study had dysphagia because of newborn juxtaoral structural problems. The infant was premature and consistently cared for at the NICU. An operation was conducted to secure the airway that was
being impacted by airway oedema. After her condition stabilized, she got treatment for dysphagia at the department of rehabilitation medicine. At the initial assessment, the infant was mainly receiving NG tube feeding, as voluntary oral feeding was difficult, and she was showing overall deterioration of oral-motor function. Muscle tone around the oral cavity was high, and the stability of the chin and cheeks was low. In particular, the infant displayed disharmony in the “sucking-swallowing-breathing” pattern, which is the most important pattern for oral feeding. Repeated disharmony in this pattern prevents a patient from maintaining appropriate breathing between sucking, as a result of which oxygen saturation levels can fall. If hypoxia repeatedly occurs, cerebral damage can occur as blood flow is reduced and, with the delay in the development of oral feeding capability, the patient can develop growth retardation [16]. Therefore, the infant in this study was in urgent need of dysphagia treatment.

Non-nutritive sucking training is one of the most frequently used methods to improve the feeding capability of preterm babies, and it has been reported to stabilize infants with unstable breathing, which helps them promptly move to oral feeding [17]. However, conducting only non-nutritive sucking training does not influence the stability of the chin and cheeks, or the stabilization of muscle tone. To address these issues and improve oral feeding capability, many researchers have implemented oral stimulation interventions [18, 19]. Oral stimulation intervention usually refers to a massage performed 15 minutes before feeding that includes activities to stimulate the cheeks, upper lip, lower lip, upper gum, lower gum, inner cheeks, lateral border of the tongue, and middle blade of the tongue by pressing those parts with fingers. In this study, a manual oral stimulation program was implemented on the infant with Pierre Robin Syndrome for three weeks. The study was structured with a single-subject research design using the AB design. During the baseline period, only non-nutritive sucking training was offered without an oral stimulation program. During the intervention period, non-nutritive sucking training and a 30-minute oral stimulation program were conducted.

The average oral feeding volume per time during the baseline period was 5.0 ± 0.0 mL, and the average feeding volume per time during the intervention period was 17.6 mL. The average volume increased by 12.6 mL during the intervention period compared to the baseline period. The average oral feeding volume during the baseline period was 5.0 ± 0.0 mL, and the average oral feeding volume during the intervention period was 35.3 ± 2.0 mL. The average oral feeding volume increased by 30.0 mL during the intervention period compared to the baseline period. In the Lee and Park [20] study, research was conducted with a preterm baby to examine the effects of an oral stimulation program, and the study results showed an increase in the weight of the infant, the daily volume of milk, and the volume of milk per time, and a decrease in the feeding time per time. The Rocha et al. [21] study also implemented an oral stimulation program for preterm babies, and the results showed that the program enhanced oral motor skills of preterm babies which led to a significant increase in weight and feeding volume of the infants.

The feeding volume during the intervention period increased more significantly when a special feeding bottle and an oral stimulation program were applied together compared to that when only a special feeding bottle was used. This result indicates that lasting effects can be achieved if an oral stimulation program is implemented every day and that applying both an oral stimulation program and a special feeding bottle leads to the best results. We could also see that the feeding volume increased when a special feeding bottle was used compared to that when an ordinary feeding bottle was used, and we believe that this provides good guideline when choosing feeding tools for infants with a cleft palate.

The feeding time slightly decreased during the intervention period compared to that of the baseline period. Kim & Kim [22] reported that it is important to assess muscle tone, posture, oral reflexes, changes of swallowing time, breathing, and other factors when assessing whether the swallowing function has been improved in an infant for who standardized assessment tools are difficult to use. A shorter swallowing time indicates a stabilization of the “sucking-swallowing-breathing” pattern. To achieve this stabilization, the movements of the chin, tongue, and lips need to be adjusted, and the capability to move and coordinate the oral cavity with a rhythmic pattern is required [23]. In this study, the infant showed an overall deterioration of oral-motor function and a high level of muscle tone around the oral cavity. However, the muscle tone got lower as treatment was carried out, and the movements of the chin, cheeks, tongue, and lips improved. As a result, by adjusting the muscle tone, improving movements of the juxtaoral parts, and stabilizing the “sucking-swallowing-breathing” pattern, we increased the oral feeding volume and reduced the swallowing time. In
addition, we prevented the aspiration or intake of air, which made the infant take in nutrients more effectively. In this study, stimulation was regularly provided at a scheduled time of the day, which allowed us to closely observe the status of the infant.

There were several limitations of this study: First, it was conducted with a single case due to the rarity of Pierre Robin syndrome; therefore, it is difficult to generalize the effects of intervention on all Pierre Robin syndrome patients. Second, we could not check the infant during the follow-up period (after the intervention period) during which the persistence of treatment effects could be studied. In this regard, follow-up research is recommended to check whether the treatment effects last after the intervention period by using the ABA research design. However, this study is significant, because there are no previous studies on dysphagia treatment for Pierre Robin syndrome. In addition, the results of this study could be confirmed with supplemental case studies that explore the effectiveness of dysphagia treatment using oral therapy intervention that can stabilize the juxtaoral muscles and improve an infant’s oral feeding capability.

5. CONCLUSION

In this study, an occupational therapist implemented a dysphagia treatment for Pierre Robin syndrome using an oral stimulation program, and this study aimed to present the effects and results of this dysphagia treatment. We set the baseline period and the intervention period using the AB design. During the intervention period, an oral stimulation program was not implemented, non-nutritive sucking training was performed using a rubber nipple, and the infant was fed with an ordinary feeding bottle. During the intervention period, non-nutritive sucking training and an oral stimulation program were applied together, and a special feeding bottle was used. We observed that the average oral feeding volume per time and the total daily oral feeding volume of the infant substantially increased during the intervention period. In addition, we observed that the feeding volume increased during the intervention period when the special feeding bottle and oral stimulation program were used together compared to that when only the special feeding bottle was used. We also observed that the oral feeding time of the infant was slightly shortened during the intervention period compared to that in the baseline period. The oral stimulation program, which was implemented before feeding, showed very positive effects such as improvement of the oral feeding capability of the infant and stabilization of the juxtaoral muscles.

CONFLICTS OF INTEREST

The author declares no conflicts of interest.

REFERENCES


